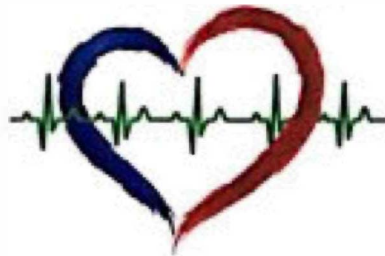


Pediatric Cardiac Intensive Care Handbook



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**The Pediatric Cardiac
Intensive Care Society**



Notice: All efforts have been made to verify that the information in this book is current at the time of publishing. Readers are advised to confirm whether new information is available about all the topics included in this book at the time of reading. Medication doses are listed in this book but ultimate dosing for individual patients must be determined by the practitioner and verified using other sources. The publishers and editors of this book assume no liability for injury related to any use of the content of this book.

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ABBREVIATIONS

AA:	arachidonic acid	CMV:	cytomegalovirus
ABG:	arterial blood gas	CO:	cardiac output
ACR:	acute cellular rejection	CO₂:	carbon dioxide
ACT:	activate clotting time	CPB:	cardiopulmonary
AMR:	antibody mediated rejection		bypass
Ao:	aorta	CPK:	creatine
aPTT:	activated partial thromboplastin time		phosphokinase
ASD:	atrial septal defect	CPR:	cardiopulmonary resuscitation
ASO:	arterial switch operation	CK-MB:	creatinine kinase-muscle/brain
AT3:	antithrombin 3	CRP:	C-reactive protein
AV node:	atrioventricular node	CT:	computerized tomography
BDG:	Bidirectional Glenn	CVL:	central venous line
BID:	twice daily	CVP:	central venous pressure
BiVAD:	biventricular assist device	CVVH:	continuous veno-venous hemofiltration
BMP:	basic metabolic panel	CXR:	chest x-ray
BNP:	brain natriuretic peptide	DCM:	dilated cardiomyopathy
BP:	blood pressure	DIC:	disseminated intravascular coagulation
BSA:	body surface area		
BT shunt:	Blalock-Taussig shunt	DO₂:	oxygen delivery
CaCl₂:	calcium chloride	DORV:	double outlet right ventricle
cAMP:	cyclic adenosine monophosphate	EBV:	Ebstein- Barr virus
cGMP:	cyclic guanosine monophosphate	ECG:	electrocardiogram
CAV:	coronary allograft vasculopathy	ECMO:	extracorporeal membrane oxygenation
CBC:	complete blood count		
CHB:	complete heart block	EDP:	end diastolic pressure
CHF:	congestive heart failure	ELSO:	Extracorporeal Life Support Organization
CI:	cardiac index	ETCO₂:	end tidal carbon dioxide

ETT:	endotracheal tube	LCA:	left coronary artery
FDA:	Food and Drug Administration	LMWH:	low molecular weight heparin
FFP:	fresh frozen plasma	LR:	lactated ringers
FiO₂:	fraction of inspired oxygen	LVAD:	left ventricular assist device
HCT:	hematocrit	LVOT:	left ventricular outflow tract
HCV:	hepatitis C virus	MAP:	mean arterial pressure
Hgb:	hemoglobin	Mcg or µg:	microgram
HIT:	heparin induced thrombocytopenia	MCT:	medium chain triglycerides
HIV:	human immunodeficiency virus	mEq:	milliequivalent
HLA:	human leukocyte antigen	mg:	milligram
HR:	heart rate	MgSO₄:	magnesium sulfate
IART:	intraartial reentrant tachycardia	Mmol:	millimol
iCa:	ionized calcium	MPA:	main pulmonary artery
iNO:	inhaled nitric oxide	MRI:	magnetic resonance imaging
INR:	international normalized ratio	NIRS:	near infrared spectroscopy
ISHLT:	International Society of Heart & Lung Transplantation	NS:	normal saline
IU:	international units	OR:	operating room
IVC:	inferior vena cava	O₂:	oxygen
IVIG:	intravenous immunoglobulins	PA:	pulmonary artery
Kcal:	kilocalories	PAC:	premature atrial contraction
KCl:	potassium chloride	PAP:	pulmonary artery pressure
Kg:	kilogram	PAPVR:	partial anomalous pulmonary venous return
L:	liter	PCR:	polymerase chain reaction
LA:	left atrium	PDA:	patent ductus arteriosus
LAD coronary:	left anterior descending	PEEP:	positive end expiratory pressure
LAP:	left atrial pressure	PFO:	patent foramen ovale
LBBB:	left bundle branch block		

PIP:	positive inspiratory pressure	SA node:	sinoatrial node
PO:	per os (oral)	SCPS:	superior cavopulmonary connection
PRA:	panel reactive antibodies	subQ:	subcutaneously
PRBC:	packed red blood cells	SVC:	superior vena cava
PTLD:	post-transplant lymphoproliferative disease	SVR:	systemic vascular resistance
PVC:	premature ventricular contraction	t_{1/2}:	half life
PVR:	pulmonary vascular resistance	TAPVR:	total anomalous pulmonary venous return
QID:	four times daily	TGA:	transposition of the great arteries
RA:	right atrium	TID:	3 times daily
RPM:	revolutions per minute	TV:	tidal volume
TOF:	tetralogy of Fallot	UFH:	unfractionated heparin
RAP:	right atrial pressure	VBG:	venous blood gas
RCA:	right coronary artery	VAD:	ventricular assist device
RR:	respiratory rate	VD:	volume of distribution
rSO₂:	regional venous oxygen saturation	VILI:	ventilator induced lung injury
RVAD:	right ventricular assist device	VO₂:	oxygen consumption
RVDC:	right ventricular dependent coronary circulation	VSD:	ventricular septal defect
		WPW:	Wolff-Parkinson-White

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FOREWORD

Pediatric cardiac critical care has emerged as a recognized and respected discipline. This handbook serves as a testament to that fact. The field can trace its origins to rapid innovations within cardiac surgery and cardiology which occurred on the heels of the polio epidemic when clinicians, scientists and managers began to think creatively about organization of care and development of specialized technology such as mechanical ventilation. With a new paradigm for care delivery of technology-dependent and high risk patients emerging, the feasibility of cardiopulmonary bypass ushered in a whole new frontier of promise and problems for children with heart disease. But the profound driver of the need for specialists and specialty care in this field was the remarkable shift in surgical techniques that allowed for early corrective and palliative surgery in neonates and young infants. It was soon recognized that these patients posed substantial new challenges in pediatric intensive care. These challenges included understanding and managing the systemic inflammatory response, the stress response, pulmonary hypertensive disorders, intricate cardiorespiratory interactions as well as attendant complications and the need for re-intervention. Solutions to these problems demanded that practitioners have a thorough and specific understanding of the nature of the circulation among infants with complex heart disease as well as expertise in treating critical illness.

Over time, we came to understand the pathophysiology underlying many of these problems, and the management during the postoperative period evolved rapidly. Indeed, this era was one of a remarkable innovation and discovery, particularly in areas related to the management of pulmonary hypertension and mechanical support of the circulation. Units dedicated to pediatric cardiac critical care evolved in large centers because of the volume, complexity and intensity of care, and from this arose a programmatic approach to the management of cardiac disease in newborns, infants and children. An integrated approach involving collaboration between cardiology, cardiac surgery, cardiac anesthesia, nursing and critical care physicians proved to be a model that enabled rapid advances in the field, the development of consistent practices and in particular, important and collaborative research that substantially improved outcomes. Initially outcomes were focused on survival, unintended complications and re-intervention. As our field evolved, however, our outcomes became much broader and related to the quality and safety of the care we deliver as well as the impact our care has on longer term neurodevelopmental outcomes and quality of life.

As the discipline became more defined in scope, so too we saw the development of new training programs in pediatric cardiac intensive care. Clinicians obtained advanced training and many completed combined board certifications in pediatric cardiology and in critical care. There is no doubt that pediatric cardiac intensive care units are integral parts of any well-functioning heart center in the current era.

Within pediatric cardiac intensive care, there has also been substantial horizontal integration, namely between physicians, nursing and allied health professionals. We have become much less hierarchical in our organizational structures and successful pediatric cardiac intensive care units have developed close partnerships and leadership between intensive care physicians, nurses and allied health professionals. Perhaps more than ever this was evident at the 10th International Conference of the Pediatric Cardiac Intensive Care Society held in Miami; attended by over 500 participants, the collaboration and energy across disciplines caring for patients with congenital heart disease was palpable. As a discipline, pediatric cardiac intensive care has grown into a recognized and important subspecialty. We have developed a body of research that continues to shape our field. Education and training opportunities across medical and nursing fields continue to expand. We have developed a knowledge base specific to the patients we care for in our environment, enabling important new initiatives related to developing consistent practices and sharing of data. This *Pediatric Cardiac Intensive Care Handbook* is the latest example of effective interdisciplinary collaboration and is an important addition to the education and training within our specialties. Written by leaders in the field and leaders at the bedside, this handbook is relevant for all of us who take care of newborns, infants and children with heart disease. The information is evidence-based, up-to-date and presented in such a way that it can be used as a real time, just in time resource. The online version with links to images is unique and enhances the breadth and depth of information about specific lesions and considerations for management. This interdisciplinary handbook is a valuable resource and an important contribution in pediatric cardiac intensive care.

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CICU Quick References

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Normal Coronary Arteries

Normal Heart

- All three segments are used consecutively to describe the sequential segmental anatomy of congenital heart defects
- Normal anatomy: {S, D, S} situs solitus atria {S}, normal D- looped ventricles {D}, and a normally committed aorta to the left ventricle {S}

Visceroatrial Situs (refers to sidedness of right atrium, liver and superior vena cava)	
{ S }	Situs solitus: right sided morphologic right atrium, SVC and liver (normal)
{ I }	Situs inversus: left sided morphologic right atrium, SVC, and liver
{ A }	Situs ambiguous: Indeterminate atrial morphology and venous connections, stomach may be midline or on either side of the abdomen, may be associated with asplenia or polysplenia and/or intestinal malrotation

Ventricular Looping (atrioventricular valves follow the morphological ventricle)	
{ D }	RV is anterior and to the right of the morphologic LV (normal)
{ L }	RV is posterior and to the left of the morphologic LV
{ X }	Ventricular position is ambiguous

Great Vessel Connection	
{ S }	Aorta is leftward and posterior to pulmonary artery (normal)
{ I }	Inverted normally related great arteries
{ D }	Aorta is anterior and to the right of the pulmonary valve
{ L }	Aorta is anterior and to the left of the pulmonary valve
{ X }	Position of the great vessels is ambiguous

Commonly Used Equations

1. Cardiac Output (CO)

$$\text{Cardiac Output (L/min)} = \text{Heart Rate (HR)} \times \text{Stroke Volume (SV)}$$

$$\text{Cardiac Index (L/min/m}^2\text{)} = \frac{\text{Cardiac output}}{\text{Body surface area}}$$

- CO the amount of blood ejected from the left ventricle every minute (L/min).
- Stroke volume is determined by
 - Preload: the amount of fill or stretch as blood distends the ventricle
 - Afterload: the resistance the ventricle must overcome to eject blood
 - Contractility: the myocardium's intrinsic ability to contract
- Stroke volume is relatively constant in infants as the immature myocardium is unable to effectively augment contractility, therefore cardiac output is more heart rate dependent

2. Arterial Oxygen Content (CaO₂)

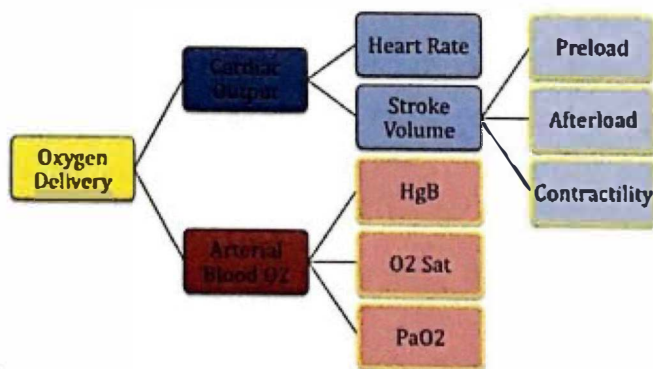
$$\text{CaO}_2 \text{ (mL O}_2\text{/dL)} = (\text{Hemoglobin} \times \text{SaO}_2 \times 1.34) + (0.003 \times \text{PaO}_2)$$

- Hgb: Hemoglobin (gm/dL)
- SaO₂: Arterial oxygen saturation
- Oxygen carrying capacity of Hb is 1.34mL/gm
- The contribution of dissolved oxygen (0.003 x PaO₂) is only 0.3 mL/dL or less. This is negligible and may be omitted (except with hyperbaric O₂ therapy)

3. Oxygen Delivery (DO₂)

$$\text{Oxygen Delivery} = \text{Arterial Blood Oxygen Content} \times \text{Cardiac Output}$$

$$\text{DO}_2 \text{ (mL/min)} = [(\text{PaO}_2 \times 0.003) + (\text{Hgb} \times 1.34 \times \text{SaO}_2)] \times \text{CO}$$



- DO_2 is the amount of oxygen delivered to the cells each minute (L/min)
- DO_2 can be compromised by severe decrease in O_2 carrying capacity (anemic hypoxia), arterial hemoglobin O_2 saturation (hypoxic hypoxia) or cardiac output (stagnant hypoxia)

4. Oxygen Consumption (VO_2)

$$\text{VO}_2 (\text{mL O}_2 / \text{min}) = \text{CO (L/min)} \times (\text{CaO}_2 (\text{mL/dL}) - \text{CvO}_2 (\text{mL/dL})) \times 10$$

- VO_2 : The rate that oxygen is removed from blood for tissue perfusion (mL O_2 /minute)
- CaO_2 : Arterial oxygen content = $(\text{Hgb} \times \text{SaO}_2 \times 1.36) + (0.003 \times \text{PaO}_2)$
- CmvO_2 : Mixed venous oxygen content = $(\text{Hgb} \times 1.36 \times \text{SvO}_2)$
- Classically mixed venous saturation (SvO_2) is measured from the pulmonary artery
- In mixing cardiac lesions, the SVC saturation is preferred
- Normal SvO_2 is 70 – 75% (in the setting of normal systemic saturations)
- VO_2 can be calculated by spirometry, by indirect calorimetry, or estimated by a nomogram

Relationship of Oxygen Delivery and Oxygen Consumption



- Typically, oxygen delivery (DO_2) increases as oxygen consumption (VO_2) increases to meet metabolic demand
- If DO_2 decreases and VO_2 remains constant, oxygen extraction increases to compensate. Once tissues become hypoxic, anaerobic metabolism ensues and lactate is produced as the byproduct, causing a metabolic acidosis
- Maintaining DO_2/VO_2 balance is critical in the management of CICU patients
- Optimizing DO_2 and minimizing VO_2 is a strategy used to maintain adequate oxygen delivery in states of imbalance

Strategies to Decrease Oxygen Consumption(VO_2)	Strategies to Increase Oxygen Delivery (DO_2)
• Positive pressure ventilation • Paralysis • Sedation • Avoid high consumption states: ○ Seizures or shivering ○ Hyperthermia ○ Infection • Avoid high dose inotropes if possible (will increase myocardial VO_2)	• Increase CaO_2: ○ Optimize hemoglobin ○ Optimize saturation • Optimize CO: ○ Ensure adequate preload ○ Assure AV synchrony ○ Inotropic support ○ Reduce afterload ○ Allow for diastolic filling time

5. Fick Equation for Cardiac Output (CO)

$$CO \text{ (L/min)} = \frac{VO_2}{(CaO_2 - CvO_2)}$$

$$\text{Simplified CO (L/min)} = \frac{VO_2}{(AO_{sat} - MV_{sat}) \times Hgb \times 1.36 \times 10}$$

- Fick principle utilizes oxygen consumption and the arterio-venous oxygen content difference to calculate cardiac output (Q). This equation can be applied to the systemic circulation (Q_s) and the pulmonary circulation (Q_p) to determine the ratio of pulmonary to systemic blood flow in patients with intracardiac shunting
- AO_{sat} : aortic saturation
- MV_{sat} : mixed venous saturation
- VO_2 is generally obtained from nomogram or estimated

$$Q_p / Q_s = \frac{AO_{sat} - MV_{sat}}{PV_{sat} - PA_{sat}}$$

- PV_{sat} : pulmonary venous saturation (assumed to be 95-99% if there is no lung disease)
- PA_{sat} : pulmonary artery saturation (equals aortic saturation (AO_{sat}) in single ventricle patients)
- Normal Q_p/Q_s ratio is 1:1
- With a left to right intracardiac shunt (e.g. large VSD), pulmonary blood flow can be much higher than systemic blood flow (i.e. a Q_p/Q_s ratio of >1:1 or more)
- With a right to left intracardiac shunt (e.g. TOF), pulmonary blood flow is less than systemic flow and Q_p/Q_s ratio <1:1

6. Arterial - Venous Oxygen Delivery Difference (A-V DO₂)

$$\text{A-V DO}_2 = \text{CaO}_2 - \text{CvO}_2$$

- A-V DO₂ is an indication of how much oxygen is removed from the blood in capillaries
- In the setting of low DO₂, oxygen extraction increases to maintain a normal VO₂. If increased oxygen extraction is insufficient to maintain VO₂, cardiac output increases to improve DO₂
- If increased cardiac output and oxygen extraction are insufficient, VO₂ will fall and DO₂ will fall below the critical DO₂
- Low A-V DO₂ may be secondary to increased oxygen extraction from arterial blood suggesting high oxygen consumption state (i.e., sepsis) or to decreased oxygen delivery (i.e., low cardiac output syndrome)

7. Alveolar - Arterial Gradient (A-a gradient)

$$\text{A-a gradient} = \text{PAO}_2 - \text{PaO}_2$$

- A-a gradient is the difference between alveolar oxygen concentration and arterial oxygen concentration
- Normal A-a gradient is 10-20
- A-a gradients are difficult to interpret in children with cardiac shunting
- PAO₂: the partial pressure of O₂ in the alveoli
- PaO₂: the partial pressure of O₂ measured in the arterial blood gas

$$\text{PAO}_2 = \text{FIO}_2 \times (\text{P}_B - \text{P}_{\text{H}_2\text{O}}) - (\text{PaCO}_2/\text{RQ})$$

- F_IO₂: (Fraction of inspired oxygen)
- P_B: Barometric pressure (760mmHg at sea level)
- P_{H₂O}: Water vapor pressure (47 mmHg at 37° C)
- RQ: Respiratory quotient=CO₂ eliminated/CO₂ consumed (0.8 usually)

$$\text{Simplified PAO}_2 (\text{at sea level}) = \text{FIO}_2 \times (713) - \frac{\text{PaCO}_2}{0.8}$$

8. Coronary Perfusion Pressure

$$\text{Coronary perfusion pressure (mmHg)} = \text{Arterial diastolic blood pressure} - \text{LVEDP}$$

Notes

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Notes

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CHAPTER 2

Routine CICU Postoperative Management

Chapter Editors

Erica Molitor-Kirsch, MD

Dawn Tucker, MSN, CPNP-AC

Section 1: OR to CICU Hand-off

Section 2: Hemodynamic Monitoring

Section 3: Respiratory Management

Section 4: Fluid and Electrolyte Management

Section 5: Nutrition Management

Section 6: Postoperative Laboratory Tests

Section 1
OR to CICU Handoff
Christine M. Riley, MSN, APRN, CPNP-AC

Importance of a Structured Handoff

- OR to CICU transition is a time of great vulnerability after congenital heart surgery
- Standardized team-based handoffs optimize accuracy and minimize lapses in care
- Essential personnel
 - OR team: anesthesiologist, surgeon
 - ICU team: attending, ICU fellow/APN/resident, bedside nurse, respiratory therapist

Components of OR to CICU Handoff

- Transfer of equipment
 - Airway and ventilator
 - Monitoring equipment
 - Lines, tubes, and drains
 - Vasoactive and other infusions
- Patient details
 - Age, weight, preoperative anatomy and physiology, comorbid conditions
- Perioperative details
 - Anesthesia induction, airway or intubation considerations, endotracheal tube size/type
 - Access: type, size, site, difficulties
- Intraoperative details
 - Cannulation and transition to cardiopulmonary bypass
 - Details of repair
 - Cardiopulmonary bypass and cross clamp times
 - Intraoperative complications
 - Transesophageal echocardiographic findings
- Postoperative details
 - Hemodynamics during separation from cardiopulmonary bypass
 - Vital signs: temperature, HR, RR, BP, O₂sats
 - Hemodynamic measurements: RAP, LAP, PAP, NIRS
 - Arrhythmias and need for defibrillation, cardioversion or temporary pacing
 - Vasoactive and sedation medications
 - Extent of bleeding and required blood products

- Ventilator support and settings
 - Last antibiotic dose and time
- Anticipated postoperative problems
 - Low cardiac output syndrome, arrhythmia, bleeding
 - Defect/repair specific concerns
- Expected Trajectory
 - Timeline for maintaining mechanical ventilation
 - Hemodynamic parameters

Section 2

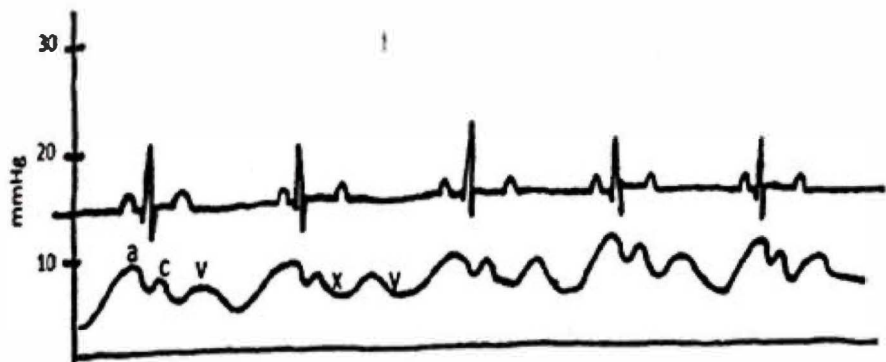
Hemodynamic Monitoring

Sherrill D. Gutierrez, MD, Jennifer Flint, MD, Dawn Tucker, MSN, CPNP-AC,
Laura Miller-Smith, MD

Intracardiac Pressure Monitoring

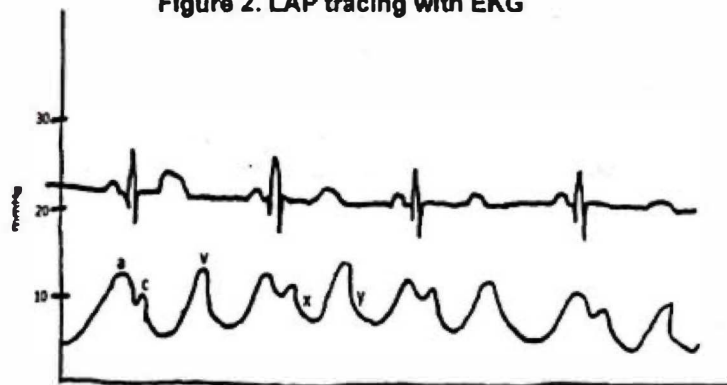
- Right Atrial (RA) Catheter
 - Tracings/Waveforms
 - a wave: atrial contraction
 - c wave: bulging of TV into RA
 - x descent: TV pulled away from RA during RV contraction
 - v wave: before opening of TV during late systolic filling of RA
 - y descent: TV opening and RV filling (Figure 1)
 - Interpretation
 - Normal RA pressure (RAP): 0-5 mm Hg
 - Reflects RV end diastolic pressure (RVEDP) in the absence of TV stenosis
 - The extent to which either ventricle fills is the product of ventricular diastolic transmural pressure (EDP – surrounding pressure) and compliance; thus the optimal filling pressure must be determined for each patient in a variety of settings (eg., spontaneous breathing vs. PPV)
 - Increased RAP: decreased RV compliance, increased RV volume, tricuspid valve stenosis
 - Decreased RAP: compliant RV, decreased RV volume

Figure 1. RAP tracing with EKG



- Left Atrial (LA) Catheter
 - Tracings/Waveforms
 - a wave: atrial contraction
 - c wave: bulging of mitral valve into LA after valve closure
 - v wave: before mitral valve opening during late systolic filling of LA
 - Interpretation
 - Normal LA pressure (LAP): 5-12 mmHg
 - Reflects LVEDP in the absence of MV stenosis
 - The extent to which either ventricle fills is the product of ventricular diastolic transmural pressure (EDP – surrounding pressure) and compliance; thus the optimal filling pressure must be determined for each patient in a variety of settings (eg., spontaneous breathing vs. PPV)
 - Increased LAP: decreased LV compliance, increased LV volume, mitral valve stenosis, loss of atrioventricular synchrony
 - Decreased LAP: compliant LV, decreased LV volume

Figure 2. LAP tracing with EKG

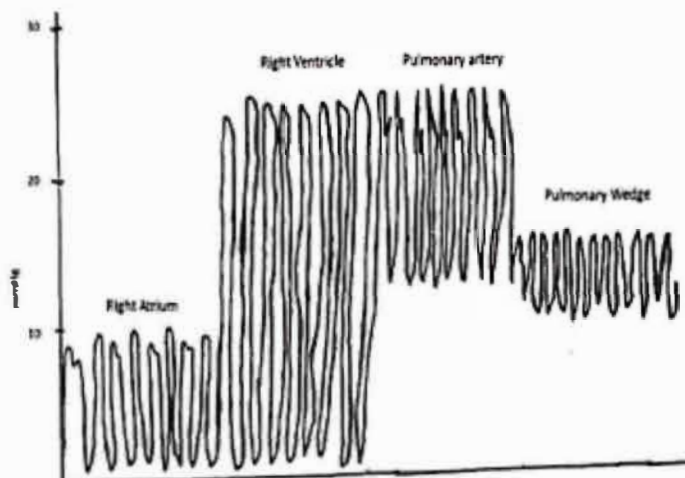


- Pulmonary Artery (PA) Catheter
 - Allows for direct measurement of RA and PA saturations and pressures, pulmonary capillary wedge pressures (PCWP), and cardiac output (CO)
 - Able to calculate systemic and pulmonary vascular resistance, stroke volume, and oxygen consumption (please see Chapter 1: CICU Quick References)
 - Cardiac output (CO)
 - Measured using thermodilution technique
 - Must know volume and temperature of injectate

- $\frac{V(TB-SI)}{A} \times \frac{(SI \cdot CI)}{(SB \cdot CB)} \times \frac{60 \cdot CT \cdot K}{1}$
- V = volume of injectate, TB = temperature of blood, A = area of thermodilution curve in square millimeters divided by paper speed (mm/sec), SI/SB = specific gravity of blood and injectate, CI/CB = specific heat of blood and injectate, CT = correction factor for injectate warming, K = calibration constant
- Cardiac index (CI) = CO / BSA
- Normal = 3.5 – 5.5 L/min/m²
- Stroke volume index = CI / HR
- Normal = 30 – 60 mL/m²
- Pulmonary vascular resistance index (PVRI) calculation
 - PVRI = [(mean PAP – PCWP) / CI] x 80
 - Normal range: 80-240 dynes ·sec/cm⁵/m²
 - Wood units: PVRI / 80 = Units/m²:
 - Normal range: 1-3 Wood Units-m²
- Systemic vascular resistance index (SVRI) calculation
 - SVRI = [(MAP - CVP) / CI] x 80
 - Normal range: 800-1600 dynes ·sec/cm⁵/m²
- Tracings/Waveforms
 - Pressures are monitored during insertion (Figure 3)
 - The catheter passes into the RA, RV and then the balloon is inflated and floated' across the pulmonary valve into the pulmonary artery, at which time an increase in the diastolic pressure occurs
 - The catheter is advanced until it is wedged
 - After wedge pressure obtained, deflate balloon and confirm restoration of PAP waveform
- Interpretation
 - PCWP: See LAP above
 - Large v wave indicates mitral regurgitation or decreased ventricular compliance
 - Cannon a waves seen in AV dyssynchrony
 - Pulmonary arterial pressures
 - Ideally measured with catheter tip at level of or below LA on lateral CXR (West Zone 3) and at end expiration
 - Normal PAP: <2/3 systemic pressure; PA systolic pressure: 15-30 mmHg; PA diastolic pressure: 5-15 mmHg
 - Normal transpulmonary pressure gradient (mean PAP – PCWP): <20 mmHg

- Pulmonary artery oxygen saturation
- True mixed venous O₂ saturation if no intracardiac shunting present
- O₂ saturation step-up of >5% from SVC to PA suggests presence of a left to right shunt

Figure 3. PA catheter tracing



Cardiac Output Monitoring

- Venous oximetry
 - Please see Chapter 1: CICU Quick References
- Near Infrared Spectroscopy (NIRS)
 - Continuous non-invasive determination of regional venous oxygen saturation (rSO₂)
 - Measures the amount of light absorbed by oxygenated and deoxygenated hemoglobin
 - The algorithm measures light absorption in nonpulsatile blood; because a majority of blood in the microcirculation is venous (75% or so) the rSO₂ is used as a surrogate for venous oxygen saturation of the interrogated viscera
 - As with pulse oximetry, which looks at the pulsatile signal, the value is calculated from an algorithm (i.e., it is not measured)
 - Because the NIRS monitor relies on an algorithm to calculate rSO₂ as well as other technical factors, the rSO₂ may vary considerably from measured values

- Interference: increased superficial tissue, melanin, bilirubin, body wall edema
- The somatic NIRS oximeter probe may be placed over the lumbar area and abdominal wall to assess renal and splanchnic tissue oxygenation. Cerebral NIRS oximeter probe may be placed on the forehead. The data on the accuracy and utility of such measurements remains to be determined
- Interpretation of cerebral rSO₂
 - Same as for venous oximetry
 - The cerebral OER assesses the cerebral O₂ supply and demand relationship and may be used to infer the global O₂ supply and demand relationship
- Lactate
 - Lactate is a biochemical indicator of the global O₂ supply and demand relationship. Lactate is a weak anion that is a byproduct of anaerobic metabolism, which produces an elevated anion gap metabolic acidosis.
 - Causes of elevated lactate
 - Type A: Shock state characterized by O₂ supply demand imbalance
 - Type B: May be caused by medications, impaired renal and hepatic clearance and by an inborn error of metabolism
 - Measurement and levels
 - Arterial and venous samples correlate well, normal levels vary by assay used
 - Levels should be measured within 15 minutes of obtaining sample
 - Treatment and prognosis
 - Elevated lactate levels and inability to clear lactate is associated with increased mortality in children after congenital heart surgery
 - Treatment should be targeted at the underlying cause and by optimizing oxygen supply and demand
- Pulse Index Continuous Cardiac Output Monitor (PiCCO)
 - Catheter placement and requirements
 - Requires PiCCO specific arterial catheter with attached thermistor and central venous catheter
 - Most commonly placed in femoral artery; may be placed in radial, brachial or axillary arteries if the vessel can accommodate the catheter

- Transpulmonary thermodilution
 - Known quantity of cold injectate is infused through the central venous catheter and the thermistor in the arterial line measures changes in temperature over time
- Very good correlation with measurements derived by pulmonary artery thermodilution and by the Fick method
- The thermodilution-derived measurement of CO allows for initial and periodic calibrations of the pulse contour-derived CO measurement
- Continuous CO measurements by pulse contour analysis
 - A technology based on the principle that the area under the systolic portion of curve of a central arterial waveform correlates with the stroke volume
 - Accuracy impacted by aortic and arterial compliance, which varies significantly among patients and disease states
 - Unreliable data in patients with intracardiac shunts or valvular lesions

Section 3
Respiratory Management
Richard Speicher, MD and Aimee Jennings, RN, CPNP

Intubation Pearls

- **Necessary equipment**
 - Flow-inflating manual ventilation bag
 - Oxygen source
 - Appropriately sized oropharyngeal airway
 - Suction catheters and suction source
 - Laryngoscope
 - Newborn 0-1 straight blade
 - <2 years: 1-2 straight blade
 - >2 years: 2+ curved blade
 - Quantitative ETCO₂ monitor
 - Endotracheal tubes (ETT) with stylet for oral intubation, cuffed tube preferred
 - ETT size = $\frac{\text{age in years} + 4}{4}$
 - ETT depth: $\frac{\text{age in years}}{2} + 12$ (at the gumline)
- **Rapid sequence intubation**
 - Indicated in patients with gastric contents or unknown NPO status to minimize risk for aspiration
 - Pre-oxygenate patient either by allowing patient to breathe spontaneously from a non-rebreathing mask and reservoir or if necessary provide bag-valve mask ventilation
 - Administer sedative, analgesic, and neuromuscular blockade in rapid sequence

Mechanical Ventilation

- Positive pressure ventilation can have significant effects on hemodynamics, particularly in children with underlying heart disease
 - Positive pressure ventilation reduces LV afterload and may be helpful with LV or systemic ventricle dysfunction
 - Spontaneous ventilation or negative pressure ventilation improves systemic venous return
 - Patients with passive pulmonary blood flow or RV systolic dysfunction may benefit from spontaneous or negative pressure ventilation due to lower RV afterload
- General conventional mechanical ventilator principles
 - Targeted tidal volumes (TV) of 8-10 mL/kg
 - In patients with acute lung injury/acute respiratory distress syndrome, low tidal volume ventilation (~6mL/kg) with peak inspiratory pressure (PIP) < 30-35cm H₂O may be used to minimize ventilator induced lung injury (VILI)

- Peak end expiratory pressure (PEEP) 4-5cm H₂O to maintain lung volume in the absence of lung and chest wall disease (e.g., abdominal distension, chest wall edema)
- Ideal PEEP in lung disease results in an adequate end expiratory lung volume while minimizing its potentially adverse impact on systemic venous return and RV afterload
- Rate/frequency should be set to age specific norms, status of lung parenchyma (e.g., pulmonary edema), and to achieve specific PaCO₂ targets, and then titrated accordingly
- Pressure support provides a set pressure to assist with patient initiated breaths and is typically set between 8-10cm H₂O
- Typical I:E ratios range from 1:2 to 1:3 and inspiratory time should be set accordingly
- FiO₂ settings will be variable based on inherent lung characteristics and considerations of cardiac anatomy and intracardiac shunting
 - Prolonged FiO₂ >0.60 should be avoided to minimize lung toxicity
- Volume Control (VC)
 - A set tidal volume is delivered resulting in variable peak inspiratory pressure (PIP), as determined by lung compliance and airway resistance; the PIP should be closely monitored to avoid barotrauma
 - Breaths are delivered with a square wave flow pattern with constant inspiratory flow
- Pressure Control (PC)
 - Delivers breaths at a constant PIP for a pre-determined cycle length based on a set inspiratory time
 - Tidal volume and minute ventilation are dependent variables determined by lung compliance and airway resistance; should be carefully monitored to avoid hypoventilation and/or hyperventilation and volutrauma
 - Breaths are delivered with a decelerating wave flow pattern
- Pressure Regulated Volume Control (PRVC)
 - A volume-targeted pressure control mode of ventilation that delivers a set tidal volume by breath to breath alterations in inspiratory pressure
 - Provides a constant inspiratory pressure with a decelerating wave flow pattern which may improve ventilation/perfusion matching and oxygenation
 - Advantages include allowing for a set minute ventilation provided the pressure limit is not reached which may limit tidal volumes
 - The ability to set a pressure limit reduces the risk for barotrauma

- **Neurally Adjusted Ventilatory Assist (NAVA)**
 - Ventilator technology which utilizes a naso-esophageal sensor to detect diaphragm electrical activity and synchronizes ventilator breaths to the patient driven inspiratory/expiratory cycle
 - Benefits of improved synchronization may include improved comfort due a faster breath trigger time, less need for sedatives, and lower PIP
 - Limitations are due to difficulty with electrical interference impairing diaphragmatic sensing
- **Airway Pressure Release Ventilation (APRV)**
 - Inverse ratio method of ventilation using continuous CPAP with brief, time-cycled, intermittent releases in pressure
 - Prolonged inspiratory time results in improved oxygenation due to higher mean airway pressures, while the release periods provide tidal volume and facilitate ventilation, while allowing spontaneous breathing
 - Benefits include improved lung recruitment, higher mean airway with lower peak airway pressures, and the ability for patients to spontaneously breath
 - Caution must be used in the post-operative cardiac patients as the higher constant mean airway pressure may increase intrathoracic pressure, thus impeding venous return
- **High Frequency Oscillatory Ventilation (HFOV)**
 - Ventilator strategy delivering very small tidal volumes at a high frequency resulting in the ability to provide increased mean airway pressure while minimizing the risk of barotrauma associated with high peak pressures
 - Increasing mean airway pressure will improve oxygenation,
 - Increasing the ΔP or decreasing the frequency will improve CO_2 removal
 - Caution must be used in the postoperative cardiac patients as the higher constant mean airway pressure may increase intrathoracic pressure, thus impeding venous return

Non-Invasive Respiratory Support

- **Continuous Positive Airway Pressure (CPAP)**
 - Provides continuous positive pressure during all phases of the respiratory cycle
 - May be helpful in patients with airway obstruction (increasing airway transmural pressure) and in maintaining lung volumes in patients with parenchymal lung disease
 - Does not synchronize with patient breaths and interface can be difficult in infants and young children

- **Bi-level Positive Airway Pressure (BiPAP)**
 - **Inspiratory pressure, an end expiratory pressure, and rate are set**
 - **Benefits of CPAP with assisted ventilation**
 - **Interface can be difficult with infants and young children**
- **High Flow Nasal Cannula (HFNC)**
 - **Provides high flow of humidified oxygen through a nasal cannula**
 - **May provide some degree of CPAP**

Section 4
Fluid and Electrolyte Management
Trina Blunk, MSN, APRN, CPNP-PC and Michael Hobson, MD

Immediate Physiologic Effects from Cardiopulmonary Bypass

- Inflammatory response: Increased capillary permeability leads to an increase in interstitial fluid
- Intravascular volume status may be dynamic and variable due to
 - Vasodilation from CPB-induced inflammation
 - Ongoing losses: chest tube output, postoperative bleeding and third-spacing
 - Impaired ventricular function
 - Impact of positive pressure on systemic venous return
- The first priority is to maintain adequate preload and cardiac output
- Fluid bolus options
 - Crystalloids: NS or LR
 - Colloids: 5% albumin
 - 5-10 mL/kg boluses
 - Theoretical advantage of raising oncotic pressure and remaining in the intravascular space for a longer duration
 - Blood products
 - PRBCs to replace active blood loss or optimize hemoglobin

Daily Fluid Requirements

- Traditional maintenance fluid requirements for non-cardiac surgery patients (4,2,1 rule)
 - 0-10 kg: 4mL/kg/hour
 - 11-20 kg: 2mL/kg/hour
 - > 20 kg: 1mL/kg/hour
- Cardiac surgery patients
 - Institution specific, consider
 - CPB cases: 50-75% maintenance on postoperative day 0, liberalize thereafter
 - Non CPB cases: 75-100% maintenance on postoperative day 0, liberalize thereafter
- Dextrose composition
 - 10% dextrose for neonates
 - 5% dextrose for infants and children
 - Tight glycemic control (i.e., targeting normoglycemia) is not supported by the findings of two multicenter randomized trials in children recovering from cardiac surgery
- Sodium composition
 - 0.45% NS for neonates
 - 0.45-0.9% NS for infants and children

Fluid Removal

- Diuretic therapy
 - Commonly instituted on postoperative day 1
 - Diuretic medications listed in pharmacologic section
- Dialysis
 - Peritoneal dialysis
 - Remove fluid
 - May aid in the removal of soluble inflammatory mediators
 - Peritoneal dialysis catheters are routinely used for passive peritoneal drainage in some centers

Electrolyte management

- Potassium
 - Goal level >3.5 mmol/L, >4 mmol/L if patient prone to dysrhythmias
 - Replacement: KCl 0.25-1 mEq/kg IV over 1-2 hours and/or add to maintenance fluid
 - Use with caution in patients renal dysfunction
- Magnesium
 - Goal level > 2 mg/dL, particularly if patient is prone to dysrhythmias
 - Replacement: MgSO₄ 20-50 mg/kg IV over 2-4 hours
 - May cause vasodilation/hypotension if given too rapidly
- Calcium
 - iCa goal level > 1 mmol/L
 - Replacement
 - CaCl₂ 10-20 mg/kg IV over 30 minutes
 - Preferably via CVL due to risk of extravasation and tissue injury
 - Calcium gluconate 60-100 mg/kg IV over 1 hour
 - Less readily bioavailable compared to CaCl₂ due to hepatic dissociation

- Zinc is important for wound healing and cardiac patients may be zinc deficient
- Low zinc levels are closely correlated with low alkaline phosphatase levels. Serum zinc levels may also be measured directly and supplemented if low
- Iron
 - Iron deficiency is also common in the congenital heart disease population
 - Serum ferritin level is the most accurate measure of iron stores

Special Feeding Concerns

- Chylous effusion
 - Assess the etiology of the chylous effusion (i.e., increased SVC pressures, innominate vein clot) and consider medical, cardiac catheterization or surgical intervention if indicated
 - Chest tube output >20mL/kg/day: consider parenteral nutrition and NPO status for 7-10 days
 - Chest tube output is <20mL/kg/day: utilize formula high in medium chain triglycerides or skimmed breast milk with added MCT oil

Section 5
Nutrition Management
Julie Slicker, MS, RD, CSP, CD, CNSC

Parenteral Nutrition (PN)

- Commonly instituted on postoperative day 2-3 if enteral feeding not possible
- Caloric goals
 - Infants 90-100 kcal/kg/day
 - Toddlers 80-90 kcal/kg/day
 - Children 45-70 kcal/kg/day
- Fluids: Please see Chapter 2 Section 4: Daily Fluid Requirements
- Dextrose: Start with Glucose infusion rate (GIR) of 4-6mg/kg/min, advance by 2-4mg/kg/day to goal
 - Infants 14-16 mg/kg/min
 - Toddlers 12 mg/kg/min
 - Children 8-10 mg/kg/min
- Amino Acids
 - Infants: 3 g/kg/day
 - Toddlers: 2-3 g/kg/day
 - Children: 1.5-2 g/kg/day
 - Limit in patients with renal dysfunction
- Lipids
 - Infants 3 g/kg/day, children 1.5-2.5 g/kg/day
 - Hypertriglyceridemia (triglycerides>300) or cholestasis: limit lipids to run over 12-16 hours and/or decrease to 1.5 g/kg/day
 - Levocarnitine 8-10 mg/kg/day may be considered to optimize lipid metabolism
- Micronutrients/trace elements
 - Standard micronutrients/trace elements are typically initiated
 - Adjustments may be required due to diuretic therapy, other medications, losses and/or physiologic status

Considerations Regarding Enteral Feeding

- Ensure adequate systemic and mesenteric perfusion prior to initiation
- Anatomic considerations: heterotaxy, maxillofacial abnormality, increased abdominal girth
- Physiologic: excessive vomiting and/or diarrhea, hematochezia, necrotizing enterocolitis

Enteral Nutrition

- Commonly instituted as early as postoperative day 1 depending on cardiorespiratory status

- **Enteral nutrition options**
 - Oral feeding: appropriate in patients with adequate cardiopulmonary function and adequate aerodigestive coordination and function
 - Nasogastric or nasojejunal tubes may be placed for nutritional support
 - Gastrostomy or jejunostomy tubes may be utilized as a more permanent means to providing nutrition support in patients with inadequate oral-motor skills
 - Bolus feeds are physiologic and more conducive for progressing oral feeds
 - Enteral feeding protocols have been shown to decrease practice variation, optimize the progression of enteral feeds and decrease complications related to parenteral nutrition
 - Studies have demonstrated that outcomes are improved the earlier enteral nutrition is initiated and the sooner goal enteral nutrition is reached and parenteral nutrition is discontinued
- **Caloric goals**
 - Infants 100-130 kcal/kg/day
 - Toddlers 90-100 kcal/kg/day
 - Children 60-80 kcal/kg/day
 - Caloric requirements are dynamic and full physical and anthropometric assessments should be done to determine individual calorie and growth needs
- **Human milk / formula selection**
 - Human milk is the ideal enteral selection for infants
 - Standard formula is appropriate for older children or when human milk is unavailable
 - Specialty formulas may be warranted if standard formulas cause intolerance
 - Formulas high in medium chain triglycerides (MCT) are useful in cases where systemic perfusion is compromised or chylous effusions are present

Micronutrient Deficiencies

- **Vitamin D**
 - The congenital heart disease population is at a particularly high risk for vitamin D deficiency due to inactivity, low nutrient reserves and high metabolic demand
 - Goal 25-OH Vitamin D level during the postoperative period is >30mg/dL
 - All infants and children should receive a minimum of 400 IU/day of cholecalciferol (Vitamin D₃)
 - In deficiency: add an additional 400 – 1000 IU daily. Consider endocrine consult
- **Zinc**

Section 6
Postoperative Laboratory Tests
Molly Lappe, MSN, RN, CCRN and Erica Molitor-Kirsch, MD

Standard Laboratory Tests Upon Arrival to CICU

- Specific labs and frequency are center specific
- ABG / VBG: evaluate oxygenation, ventilation, and acid-base balance
- Lactate: elevated in the setting of inadequate oxygen delivery
- CBC with differential
- Coagulation panel
- Basic metabolic panel
- Ionized calcium, magnesium, and phosphorus
- Albumin
- 12 lead ECG to have baseline parameters by which to compare future studies

Routinely Scheduled Postoperative Labs

- Patient and center dependent
 - Typically obtained more frequently after complex neonatal surgeries than in less complex populations
 - Routine postoperative order sets may be useful to assure standardization
- ABG: Q2-6 hours
- VBG: Q2-6 hours
- Lactate: Q2-6 hours
- CBC Q6-24 hour, differential daily
- Coagulation panel: on admission only for most cases; more frequently if bleeding or clotting concerns
- BMP: Q12-24 hours
- Ionized calcium: Q2-6 hour
 - Consider in the setting of hypotension after blood product administration
- Mg/Phos: Q day, or if arrhythmia present
- Albumin Q day (complex cases)
- Liver function test Q 24-72 hours

Additional considerations in select patients

- Thromboelastograph (TEG)

CHAPTER 3

Common CICU Postoperative Problems

Chapter Editors

Michael Gaies, MD, MPH

Catherine Baxter, MSN, CPNP-AC

Section 1: Cardiopulmonary Bypass

Section 2: Low Cardiac Output Syndrome

Section 3: Impaired Gas Exchange

Section 4: Pulmonary Hypertension

Section 5: Bleeding and Clotting

Section 6: Arrhythmia

Section 1
Cardiopulmonary Bypass
Javier Lasa, MD, FAAP

Common Techniques

- Cardiopulmonary bypass (CPB): the use of extracorporeal circulatory support to maintain oxygen delivery to all tissues during surgical procedures requiring exposure and isolation of the heart
- Cross-clamp: isolation of the heart from the systemic circulation (CPB circuit) by clamping the ascending aorta above the coronary arteries
 - Cardioplegia is instilled into the coronary arteries for myocardial protection
- Deep hypothermic circulatory arrest (DHCA): total circulatory arrest with cessation of CPB flow and core hypothermia less than 18°C
 - Allows surgeon to operate on bloodless and cannula-free field
 - Current uses include aortic arch repair or reconstruction, intracardiac repair in infants < 2-2.5 kg, and total anomalous pulmonary venous connection repair
 - Concern remains for effects on brain and subsequent neurodevelopment
- Regional cerebral perfusion (RCP): use of low flow bypass (50 ml/kg/min) to provide cerebral perfusion only via carotid artery cannula/innominate artery graft anastomosis
 - Used as an alternative to DHCA in some operations
- Ultrafiltration: use of a hemofilter to remove fluid and inflammatory mediators from circuit during or after CPB
 - Modified ultrafiltration (MUF): ultrafiltration after weaning from CPB
 - Flow is reversed, blood is drawn from arterial cannula and returned to right atrium
 - Reported benefits: removes excess total body water, reduces transfusion requirements postoperatively, attenuates coagulopathy and reduces systemic inflammatory response
 - Disadvantages include increased CPB time and complexity of the circuit

Components and Equipment

- Pump: machine that circulates circuit volume replacing native cardiac function
 - Roller pump: continuous, non-pulsatile forward flow
 - Centrifugal pumps: commonly used for ECMO and increasingly in the operating room; blood is entrained against spinning impellers (curved blades) or by creating a vortex utilizing a centrifugal cone
 - Flow is afterload dependent
- Prime: Crystalloid solution instilled in CPB circuit tubing
 - Solution contains variable amounts of electrolytes, buffer (sodium bicarbonate or THAM), calcium, and glucose depending on surgical preference

- Most priming solutions contain low levels of calcium to prevent myocardial injury during cooling
- Other potential additives include mannitol, furosemide, and methylprednisone.
- Some centers will add a colloid such as albumin, hydroxyethyl starch, or dextrans
- Red blood cells and fresh frozen plasma usually added in small patients to achieve goal hematocrit of 25% - 35%
- **Oxygenator: gas exchanger**
 - Bubble: early model oxygenator, diffuses ambient or concentrated gas into blood; least efficient oxygenator, requires large surface area
 - Silicone: permeable membrane with gas exchange occurring during countercurrent gas/blood flow; improved efficiency but susceptible to acute failure due to accumulation of blood products within membrane
 - Hollow Fiber Membrane: most commonly used, microporous hollow fibers minimize contact between gas and blood, with improved efficiency in gas exchange, lower priming volume, less foreign surface contact with lower pressure drop, and highly efficient heat exchange system
- **Cannulation techniques vary between centers and operative needs**
 - Venous sites: bi-caval, single right atrium/right atrial appendage, jugular, and/or femoral
 - Arterial sites: ascending aorta proximal to the take-off of the great vessels or femoral artery

Adverse Effects of Cardiopulmonary Bypass

- **Hemodilution decreases hematocrit, clotting factors, and plasma proteins**
 - Effects: interstitial edema, electrolyte imbalances, stress hormone release, and dysfunction of the coagulation system
 - Coagulopathy exacerbated in neonates due to immature hepatic function and diminished vitamin K-dependent clotting factors
- **Hypothermia: physiologic derangements are indirectly proportional to temperature (DHCA producing more severe effects at lower temperature)**
 - Hemodynamics: bradycardia, increased systemic vascular resistance with increased in venous return and decreased cardiac output
 - Hematologic: platelet dysfunction and altered coagulation profile
 - Renal/electrolyte: cold-induced diuresis, hypokalemia, hypophosphatemia, and hypocalcemia
 - Endocrine: decreased insulin production and function, insulin resistance, and subsequent hyperglycemia
 - Loss of cerebral blood flow autoregulation < 22 °C
- **Inflammatory response**
 - Multifaceted, multiple mediator pathways including complement, cytokines, arachadonic acid, and coagulation systems
 - Can produce acute lung injury, pulmonary hypertension, edema, coagulopathy, renal and cardiac dysfunction

Neonatal Considerations

- **Pulmonary injury: altered gas exchange**
 - The immature neonate/infant lung is predisposed to increased pulmonary vascular bed reactivity, parenchymal edema, and pulmonary hypertension after CPB
 - Diminished fat and carbohydrate reserves coupled with higher metabolic rate and oxygen consumption in neonates account for rapid hypoxia
 - Intra/extra-cardiac shunting can exacerbate the relative flow distribution between systemic and pulmonary vascular beds
- **Renal: elevated vascular resistance in neonates/infants results in limited acid-base balance control, sodium reabsorption, excretion, and dilutional capabilities exacerbated by post-CPB acute kidney injury**
- **Central nervous system: brain maturation is delayed in infants with complex CHD compared to similarly gestational age-matched neonates which increases the risk of ischemia-reperfusion injury**
- **Immune system: immature neonatal/infant immune defenses result in dysfunctional mononuclear cells, and poor antibody/complement generation post-CPB**

Section 2

Low Cardiac Output Syndrome

Jana Norris, MSN, CNS, CNP and Aparna Hoskote, MD, MRCP

Definition

- Low cardiac output syndrome (LCOS) is a transient decrease in systemic perfusion resulting in an imbalance between oxygen delivery and oxygen consumption
- Cardiac output=stroke volume x heart rate (HR). Stroke volume is dependent on preload, afterload, and contractility
- Typically occurs between 6-18 hours after cardiac surgery
- Anticipation of LCOS can aid in diagnosis and prompt treatment

Causes

- Alterations in HR or rhythm
- Decreased preload
 - Hypovolemia
 - Hemorrhage/occult bleeding
 - Diuresis
 - Inadequate fluid replacement
 - Pericardial tamponade
- Increased afterload
 - Peripheral vasoconstriction
 - Pulmonary hypertension
- Decreased contractility
 - Acidosis
 - Electrolyte imbalance
 - Myocardial injury resulting from ventriculotomy, hypoxia, or ischemia
 - Factors related to cardiopulmonary bypass
 - activation of inflammatory cascade
 - alterations in SVR and PVR
 - hypothermia
 - aortic cross clamping
 - Systolic dysfunction: decreased myocardial contractility leading to decreased ejection during systole
- Diastolic dysfunction: impaired ventricular relaxation leading to decreased ventricular compliance and elevated filling pressures
- Residual surgical abnormalities (i.e., VSD, valvular stenosis or insufficiency)

Presentation

- Poor peripheral perfusion
 - Delayed capillary refill
 - Cool extremities
 - Mottled skin
- Oliguria
- Metabolic acidosis

- Elevated lactate
- Low mixed venous saturation
- Hypotension
- Tachycardia
- Signs of cardiac tamponade

Management

- Decrease oxygen consumption
 - Sedation
 - Consider neuromuscular blockade
 - Maintain normothermia
- Optimize preload
 - Tachycardia with low CVP, low BP and low SvO₂, consider a fluid bolus
 - Assess cause for hypovolemia (i.e., bleeding, capillary leak)
- Decrease afterload
 - High SVR can impair CO. There may be compensatory increase in SVR to maintain BP as CO falls. Consider afterload reducing agents (Milrinone or Nitroprusside)
- Augment ventricular contractility
 - Inotropic support with Dopamine or Epinephrine
- Optimize ventilator management while considering cardiopulmonary interactions
 - Positive pressure ventilation reduces LV afterload and may be helpful with LV or systemic ventricle dysfunction
 - Spontaneous ventilation or negative pressure ventilation improves systemic venous return
 - Patients with passive pulmonary blood flow or RV systolic dysfunction may benefit from spontaneous or negative pressure ventilation due to lower RV afterload
- Assess underlying rhythm and treat arrhythmias
 - See Chapter 3 Section 6 for management of arrhythmias
 - Primary goal of pacing is restoration of atrioventricular synchrony
- Diastolic dysfunction: minimize tachycardia to increase diastolic filling time and decrease myocardial oxygen demands
- If LCOS persists, consider alternative therapies
 - Systemic hypothermia (35 -35.5 C) to reduce oxygen consumption
 - Stress dose hydrocortisone
 - Levosimendan, to provide inotropic effect. It also decreases SVR and PVR and has been used in postoperative LCOS. It has a long duration of action and is contraindicated in the presence of arrhythmias
- Vasodilatory shock: consider low dose Vasopressin or Norepinephrine, provided good systemic ventricular function. Vasopressors are contraindicated if poor ventricular function

Surgical Strategies if LCOS is Refractory to Medical Treatment

- Re-open chest, if not already open
- Selected children may benefit from a right to left shunt, usually at atrial level, to maintain adequate CO (i.e., RV dysfunction after Tetralogy repair, elevated transpulmonary gradient after a non-fenestrated Fontan procedure)

Extracorporeal Membrane Oxygenation

- Indications: cardiac arrest, refractory LCOS, arrhythmia, pulmonary hypertension and evidence of worsening end-organ perfusion
- Contraindications: irreversible neurological or pulmonary dysfunction and cardiac failure in patients ineligible for cardiac transplantation
- See Mechanical Circulatory Support Chapter 14 Section 1 for additional information on ECMO support

Other Supportive Measures

- Renal support
 - Peritoneal dialysis or CVVH
 - Electrolyte management
 - Maintain adequate ionized calcium, serum potassium and magnesium levels
 - Correct acid-base imbalance
- Maintain adequate hematocrit (higher in children with single ventricle physiology)
- Minimize oxygen consumption, decrease basal metabolism using adequate sedation and analgesia and control thermogenesis avoiding fever

Section 3
Impaired Gas Exchange
Louis Callow MSN, CPNP and Alissa Scheffer, MS, CPNP

Impaired Gas Exchange

- Definition: physiologic state evidenced by impaired oxygen uptake and/or carbon dioxide elimination
- Gas exchange occurs at the alveolar capillary membrane
- Efficiency of gas exchange is affected by ventilation and perfusion
- Impaired gas exchange occurs when there is a ventilation/perfusion (V/Q) mismatch

Etiology of Impaired Gas Exchange

- Decreased pulmonary blood flow
 - Anatomic defect: right ventricular outflow tract obstruction, obstruction, stenosis or hypoplasia of the pulmonary valve or pulmonary arteries
 - Increased pulmonary vascular resistance (PVR)
 - Decreased cardiac output
 - Right to left intracardiac shunt
- Pulmonary venous desaturation
 - Atelectasis
 - Anatomic defect: abnormal pulmonary venous return, pulmonary arteriovenous malformation
 - Parenchymal lung injury: chronic lung changes, acute respiratory distress syndrome
 - Pulmonary edema
 - Pleural effusion
 - Pneumonia
 - Pneumothorax
- Other factors affecting gas exchange
 - Hypoventilation
 - Airway obstruction
 - Vocal cord dysfunction
 - Anatomic airway abnormalities/compression
 - Bronchospasm
 - Tracheobronchial secretions
 - Phrenic nerve injury with hemidiaphragm paresis/paralysis
 - Patient-ventilator asynchrony
 - Increased abdominal distention/ascites/pressure
 - Mechanical/equipment: dislodgement or obstruction of endotracheal tube, ETT leak
 - Preterm infants: surfactant deficiency, abnormalities of pulmonary vasculature

Manifestations of Impaired Gas Exchange: Physical Exam

- Increased work of breathing/respiratory distress: tachypnea, retractions, nasal flaring, accessory muscle use, grunting
- Altered breath sounds: diminished breath sounds, rales, rhonchi, wheezing
- Color: cyanosis, pallor
- Altered neurological state: fatigue, decreased level of consciousness, agitation

Diagnostic Evaluation

- Laboratory: blood gas analysis, hematocrit
- Imaging: chest x-ray, echocardiogram, ultrasound, MRI, chest CT
- Invasive procedures: flexible or rigid bronchoscopy, cardiac catheterization
- Monitoring: ETCO₂, pulse oximetry, peak flow meter, oxygen consumption (VO₂) measurement

Management of Impaired Gas Exchange (see Mechanical Support section)

- Pharmacological Interventions
 - Initiation of antibiotic therapy (tracheitis, pneumonia)
 - Increase in diuretic therapy (pulmonary edema, pleural effusion)
 - Steroids (ARDS, airway inflammation)
 - Respiratory treatments: bronchodilators (bronchospasm, atelectasis), Pulmozyme (lobar collapse, thick secretions)
 - Titration of sedation/analgesia (hypoventilation secondary to pain or decreased ventilatory effort, patient-ventilator asynchrony)
 - Inhaled Nitric Oxide, Sildenafil, Illoprost, Bosentan, Flolan (increased PVR)
 - Neuromuscular blockade to decrease O₂ demand (increased pulmonary vascular resistance, patient-ventilator asynchrony)
 - Congestive heart failure management
- Respiratory interventions
 - Chest physiotherapy
 - Incentive spirometry, bubbles
 - Cough and deep breathe
 - Nasopharyngeal suction
 - Ambulation
 - Adjust ETT (displacement) or replace ETT with larger ETT (leak)
 - Oxygen (pneumothorax, increased pulmonary vascular resistance)
 - Optimize ventilatory support
- Procedural interventions
 - Pleurocentesis (pleural effusion, pneumothorax)
 - Placement of pleural chest tube (pleural effusion, pneumothorax)
 - Peritoneal drain (significant abdominal ascites affecting lung expansion)
- Additional interventions
 - Transfuse PRBC (to optimize oxygen carrying capacity, anemia)

- Optimize nutrition (hypoventilation secondary to neuromuscular weakness)
- Maintain chest tubes (if present) to suction with occlusive dressing (pneumothorax)
- Optimize positioning: elevate head of bed, prone as tolerated, position with impaired lung up (atelectasis)
- Implement ventilator associated pneumonia precautions
- Consults to pediatric specialty services: Pulmonary, Otolaryngology

Section 4

Pulmonary Hypertension

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Definition

- Elevation in pulmonary artery pressures (PAP) and pulmonary vascular resistance (PVR)
- Systolic pulmonary arterial pressure > 35 mm Hg or a mean PAP > 25mm Hg
- PAP is often compared to systemic blood pressure; if the systolic PAP is > ½ the systemic blood pressure, the patient is at higher risk for a pulmonary hypertensive event/crisis

Pathophysiology

- Endothelial cell dysfunction: increased vascular tone and pulmonary artery vasoconstriction
- Secondary increased RV pressure resulting in decreased RV systolic and diastolic function
- Decreased RV function can result in low cardiac output syndrome. Presence of an atrial septal defect or patent foramen ovale may allow a right-to-left shunt that maintains cardiac output at the expense of hypoxia

Triggering Events

- Cardiopulmonary bypass can lead to increased PVR via numerous mechanisms including
 - Endothelial injury and decreased nitric oxide (NO) production
 - Increased inflammatory response
 - Excess thromboxane production
 - Atelectasis and edema
- Hypoxia
- Acidosis (metabolic and respiratory)

At Risk Populations

- Congenital heart disease
 - Highest risk: cyanotic lesions with increased pulmonary blood flow at high pressure (i.e. truncus arteriosus).
 - Lesions with increased pulmonary blood flow at high pressure without cyanosis (i.e. unrestrictive VSD, large PDA, aortopulmonary window)
 - Pulmonary venous outflow obstruction (i.e. mitral stenosis, pulmonary vein obstruction with or without anomalous connection (TAPVR), cor triatriatum)
- Neonates in first 2-6 weeks of life who are still transitioning from fetal circulation
- Ex-premature infants with bronchopulmonary dysplasia
- Children with abnormal pulmonary reactivity (i.e. Trisomy 21)

- Respiratory disorders: alveolar hypoxia ($\text{PaO}_2 < 60\text{mmHg}$) leads to hypoxic pulmonary vasoconstriction, endothelial injury and dysfunction. (i.e. acute and parenchymal lung injury, presence of airway or respiratory center disorders, deformities of the chest wall, RSV bronchiolitis)

Presentation

- Initial signs of pulmonary hypertensive event
 - Tachycardia, hypotension, decreased perfusion, stable or decreased oxygen saturation, increased RAP, normal lactate levels and arterial BP
- Signs of pulmonary hypertensive crisis
 - Low cardiac output if no atrial communication, bradycardia, hypotension, decreased oxygen saturations, acute pallor, elevated RAP, decreased LAP, rising lactate levels
 - Worsening cyanosis if atrial communication is present
 - Can rapidly progress to cardiac arrest if not recognized

Diagnosis

- Bedside monitoring
 - ETCO_2 monitoring: increase in arterial to ETCO_2 monitoring gradient could signal decrease in effective pulmonary blood flow (i.e. a drop in ETCO_2)
 - Right atrial pressure may increase if PA pressures acutely increase
 - Decreased LAP may indicate decreased pulmonary venous return seen in acute pulmonary hypertensive crisis
 - Increased LAP may indicate decreased LV compliance (potential cause of pulmonary hypertension)
 - Pulmonary artery pressure (PAP) is the most reliable measurement of pulmonary hypertension and response to therapies
- Echocardiogram can measure PAP by Doppler interrogation of tricuspid regurgitation velocity and position of ventricular septum
- Cardiac catheterization identifies degree of pulmonary hypertension and response to therapies

Treatment

- Reduce sympathetic stimulation
 - Provide deep sedation and analgesia, consider muscle relaxants
 - Hyperoxygenate prior to and during endotracheal suctioning
 - Sedate before noxious stimulation such as endotracheal suctioning
 - Maintain normothermia
- Lower PVR
 - Maintain functional residual capacity. Avoid hyperinflation and hypoinflation of the lungs by use of adequate PEEP.
 - Maintain normal alveolar oxygen tension with oxygen
 - Although hypoxia raises PVR, hyperoxia does not lower PVR beyond what is achieved with normoxia

- Target alkalosis by ventilator manipulation or sodium bicarbonate/base infusion only in emergency
- Minimize intrathoracic pressure
- Administer pulmonary vasodilating agents
 - Inhaled nitric oxide (iNO)
 - Caution in the use of iNO in setting of left-sided obstructive lesions where increasing pulmonary blood flow and pulmonary venous return may lead to pulmonary edema without improving cardiac output
 - Rebound pulmonary hypertension may occur when discontinuing therapy may need transition to enteral pulmonary vasodilator (i.e. sildenafil)
 - Some centers follow methemoglobin levels while on iNO
 - Nonspecific agents
 - Nitroprusside
 - Milrinone
 - Sildenafil
 - Isoproterenol
 - Inhaled prostacyclin (monitor for systemic hypotension)
- Support of RV function
 - Afterload reduction with Milrinone
 - Inotropy with dopamine or low dose epinephrine
 - Vasopressors for refractory systemic hypotension to maintain systemic diastolic pressure and coronary perfusion

Section 5
Bleeding and Clotting
Michael Wolf, MD

Laboratory Tests Used for Coagulation

- Complete blood count (CBC)
 - Hemoglobin and hematocrit levels
 - Platelet count
- Protime (PT) and international normalized ratio (INR)
 - Extrinsic coagulation system (Factor VII)
 - Primary warfarin effect
- Activated partial thromboplastin time (aPTT) and activated clotting time (ACT)
 - Intrinsic coagulation system (Factors VIII, IX, XI, XII)
 - Primary heparin effect
- Fibrinogen level
 - Effect of fibrinogen on thrombin formation
- Thromboelastogram (TEG)
 - Measure of multiple factor effects on clot formation
 - Time to clot formation
 - Heparin effect
 - Clot strength
 - Fibrin formation
 - Clot lysis

Etiology of Hemorrhagic State

- Postoperative hemorrhage after cardiopulmonary bypass (CPB)
 - Heparin effect
 - Platelet dysfunction (even in setting of normal quantitative platelet count)
 - Extensive suture lines
 - Fibrinolysis (specifically in neonates)
- Hemorrhage in setting of sepsis or systemic inflammatory response syndrome (SIRS)
 - Disseminated intravascular coagulation (DIC)
 - Fibrinolysis
 - Platelet consumption
 - Clotting factor consumption

Treatment of hemorrhage

- Common blood products

Product	Content	Indication	Dosing	Notes
Cell saver: salvaged blood	Red blood cells (HCT 40-60%)	Replace blood loss	10-20 mL/kg	Autologous blood
Packed red blood cells	Red blood cells (HCT 70%)	Replace blood loss	10-20 mL/kg	
Platelets	Human platelets	Ongoing blood loss	10-20 mL/kg	Should be given even in setting of normal platelet count
Fresh-frozen plasma	All coagulation factor	Ongoing blood loss with abnormal coagulation studies	10-20 mL/kg	
Cryoprecipitate	Fibrinogen, factors VIII & XIII, vWF	Ongoing blood loss with fibrinogen level <150 mg/dL	1-2 units/5 kg body weight	

- Advanced procoagulant therapies
 - Recombinant factor VIIa
 - Indication: severe ongoing postoperative bleeding after repletion of all other coagulation products, +/- documented factor VII deficiency
 - Dose 45-90 mcg/kg
 - Adverse reactions to factor VII: increased clotting risk, allergic reaction, hemodynamic instability, DIC
 - Aminocaproic acid (Amicar)
 - Indication: severe ongoing postoperative bleeding including during ECMO
 - Mechanism: antifibrinolytic, inhibits plasmin activity
 - Dosing: bolus: 100 mg/kg, infusion: 25-35 mg/kg/hour

Hemorrhage on ECMO

- Center specific anticoagulation protocols
 - Unfractionated heparin infusion
 - Some target ACT (160-230 sec)
 - Some target anti-Xa levels (0.3-0.7 IU/ml)
- Goals of therapy for ongoing bleeding
 - HCT >35-40%

- Platelet count >100,000
- PT/INR in normal range
- Fibrinogen >150 mg/dL
- Ongoing hemorrhage despite transfusion
 - Amicar infusion
 - Recombinant factor VIIa
 - Risk of potential circuit thrombosis
 - Consider preparation of replacement ECMO circuit
 - Exploration for surgical site bleeding

Approach to Acute Postoperative Hemorrhage

- Acute postoperative bleeding
 - 10 ml/kg/hour in the 1st postoperative hour
 - 5 ml/kg/hour after 2 hours postoperatively
- Ongoing bleeding should be treated medically first unless patient is unstable
 - Replace ongoing losses with PRBCs and factors as above
 - When replacing products with severe ongoing hemorrhage
 - Ratio of blood products delivered (4:1:1 rule): for every 4 units packed red blood cells: 1 unit platelets and 1 unit fresh frozen plasma
 - Citrate in blood products can rapidly deplete calcium levels
 - Replete calcium to maintain normal ionized calcium levels
 - Consider TEG for severe ongoing bleeding
 - Can help pinpoint specific deficiency in coagulation system
 - Considerations for refractory postoperative bleeding
 - Protamine to reverse heparin effect
 - Recombinant factor VIIa
- When bleeding has been maximally medically treated surgical exploration is indicated

Acute Hemorrhagic Tamponade

- If chest tube output acutely stops draining, should raise concern for acute tamponade, especially with hemodynamic changes
- Hemodynamic signs of tamponade
 - Tachycardia
 - Hypotension often with a narrow pulse pressure
 - Elevated or rising CVP
 - Pulsus paradoxus on arterial line (or plethysmograph) tracing
- Echocardiographic signs of tamponade
 - Right atrial wall diastolic collapse
 - Mitral valve inflow variation > 25%
- Immediate interventions
 - Call the surgeon
 - Push volume: crystalloid, colloid, blood products
 - Manipulation of chest tubes to maintain their patency
 - Emergent surgical intervention

- If open chest considering open patch to decompress the pericardium
- If closed chest consider surgical exploration to drain the pericardium

Evaluation of Prothrombotic Patient

- Consider hematology consult
 - Eliminate extrinsic factors (long term indwelling central lines)
- Laboratory evaluation (most common tests)
 - Factor V Leiden
 - Prothrombin gene mutation
 - Protein C or S deficiency
 - Anti-thrombin III
 - Homocysteinuria
 - Abnormally high coagulation factors (Factor VIII)

Anticoagulation in the CICU

- At risk populations for thrombosis
 - Infants with synthetic shunts
 - All anticoagulation is center specific and variable
 - Potential treatments
 - Heparin with or without monitoring of heparin effect until tolerating enteral feeds
 - Aspirin 3-5mg/kg/day daily once tolerating enteral feeds
 - Aspirin use before full enteral feeds is also variable and center specific
 - Mechanical prosthetic valves
 - Therapeutic heparin until Warfarin is therapeutic
 - Anti-Xa level (0.35-0.7 IU/ml)
 - Warfarin for long term anticoagulation
 - Aortic or pulmonary valve: INR goal typically 2.0-3.0
 - Mitral or tricuspid valve: INR goal typically 2.5-3.5
 - Tissue prosthetic valves
 - Use of Aspirin is variable and center specific
 - Heart failure with severe ventricular dysfunction and no evidence of intracardiac thrombus
 - Aspirin 81 mg daily (if tolerating enteral feeds)
 - Heparin 10 units/kg/hour (if not tolerating enteral feeds)
 - Enoxaparin prophylaxis
 - Infants < 2 months: 0.75 mg/kg/dose Q12hours
 - Children > 2 months: 0.5 mg/kg/dose Q12hours
 - Heart failure with severe ventricular dysfunction and evidence of intracardiac thrombus
 - Warfarin (if tolerating enteral feeds). Goal INR 2-3.5
 - Enoxaparin
 - Infants < 2 months: 1.5 mg/kg/dose Q12hours

- Children >2 months: 1 mg/kg/dose Q12hours
- Goal anti-Xa level 0.5-1 IU/mL
- Therapeutic heparin infusion (if not tolerating enteral feeds). Goal anti-Xa 0.35-0.7 U/mL
 - Enoxaparin
 - Infants < 2 months: 1.5 mg/kg/dose Q12hours
 - Children >2 months: 1 mg/kg/dose Q12hours
 - Goal anti-Xa level 0.5-1 U/ml
- Left ventricular noncompaction
 - Aspirin 81 mg daily once tolerating enteral feeds
- Long term indwelling central venous lines
 - Practices vary across centers with no significant evidence supporting anticoagulation use in this setting

Section 6

Arrhythmia

Akash Patel, MD and Christopher Jordan, MD, USA

EKG Analysis of Tachycardia

- QRS is abnormal or normal (or unchanged from baseline)
 - Neonates may not have QRS > 120 milliseconds due to less ventricular mass
 - Postoperative patients often have ventricular conduction delay, QRS should be compared to baseline
- Regular or irregular rhythm
- P waves present
- P wave relationship to the QRS

Paroxysmal Supraventricular Tachycardia (SVT)

- Definition
 - Regular, typically narrow complex tachycardia
 - P waves if visible 1:1 relationship to QRS, often retrograde P waves follow QRS
 - Sudden onset and termination (often initiated by a PAC)
 - Wide QRS seen if aberrant conduction
- Additional monitor findings
 - Decreased amplitude of arterial line or pulse oximetry tracing
 - Loss of A wave with increase in CVP
- Acute treatment
 - Vagal maneuvers
 - Rapid atrial pacing (using temporary or transesophageal pacing leads)
 - Adenosine (0.1-0.2 mg/kg rapid IV push)
 - Esmolol, procainamide, amiodarone, or digoxin for incessant or recurrent SVT
 - Synchronized cardioversion for unstable SVT
- Chronic treatment
 - Digoxin or beta-blocker
 - Flecainide or Sotalol
- Pearls
 - Digoxin is contraindicated in setting of pre-excitation (Wolff-Parkinson-White (i.e. WPW))

Junctional Ectopic Tachycardia (JET)

- Definition
 - Regular, narrow QRS tachycardia
 - P waves typically dissociated from QRS with ventricular rate > atrial rate

- Occasionally can have 1:1 P wave to QRS relationship with retrograde P wave
- This pattern could also be sinus tachycardia with 1st degree AV block, overdrive pacing can distinguish
- Enhanced automaticity of AV node or His bundle usually seen in postoperative patients
- Additional monitor findings
 - CVP displays cannon A waves
- Acute treatment
 - Goal is to restore AV synchrony to maintain adequate cardiac output
 - Rate control
 - Decrease catecholamines by weaning inotropes
 - Control fever and pain
 - Therapeutic cooling
 - Sedation and paralysis
 - Correct electrolyte and acid-base abnormalities
 - Procainamide or amiodarone
 - Atrial overdrive pacing at a rate exceeding JET for AV synchrony
- Chronic treatment
 - None usually required as arrhythmia transient lasting 48-72 hours
- Pearls
 - Use atrial temporary leads to differentiate JET from other narrow complex tachycardias

Atrial Flutter

- Definition
 - Regular, reentrant atrial arrhythmia with normal QRS
 - Variable AV conduction (1:1, 2:1, 3:1 etc.); P waves related to QRS but may be > 1:1
 - Classic "saw tooth" flutter waves in the inferior leads II, III, AVF
 - May be difficult to see, adenosine may reveal flutter waves
 - Most commonly missed tachycardia in the ICU
- Additional monitor findings
 - Decreased area under curve of arterial line tracing +/- hypotension
- Acute treatment
 - Rate control: beta-blocker (i.e. esmolol), calcium channel blocker (i.e. diltiazem) if over 1 year, or digoxin
 - Rhythm control: rapid atrial pacing and/or synchronized cardioversion (0.5-2J/kg)
- Chronic treatment: uncommon, in structurally normal hearts
- Pearls
 - Adenosine can be used to block AV conduction, unmasking flutter waves
 - Consider the possibility of intracardiac thrombus prior to cardioversion if >48 hours.

- Intraatrial reentrant tachycardia (IART) is a type of reentrant atrial tachycardia which does not have the "saw tooth" flutter waves. More commonly seen in post-operative congenital heart disease.

Non-sustained Ventricular Tachycardia (VT)

- Definition
 - 3 or more consecutive PVCs with HR >120 or >20% above the upper limits of normal for age
 - P waves dissociated or not visible
 - Last <30 seconds without hemodynamic compromise
- Additional monitor findings
 - Decreased area under curve of arterial line tracing +/- hypotension
 - Cannon A waves if VT with retrograde conduction
- Acute treatment
 - Correct reversible causes (electrolytes, acidosis, fever, central line position)
 - Lidocaine or amiodarone IV
- Chronic treatment: beta-blockers, sodium channel blockers, amiodarone
- Pearls
 - Caution when using antiarrhythmics as can be pro-arrhythmic

Stable Ventricular Tachycardia

- Definition
 - Regular wide monomorphic QRS complex tachycardia
 - No visible P waves or P waves with VA dissociation
 - Can be difficult to differentiate from SVT with aberrancy or sinus rhythm with underlying bundle branch block
- Additional monitor findings
 - Decreased area under curve of arterial line tracing +/- hypotension
 - Cannon A waves if VT with retrograde conduction
- Acute treatment
 - If hemodynamically stable, correct reversible causes (electrolytes, lines, acidosis, fever)
 - Lidocaine, procainamide, beta-blocker, or amiodarone
 - If hemodynamically unstable, synchronized cardioversion with 0.5 – 2 J/kg
- Chronic treatment: beta-blockers, sodium channel blockers, amiodarone
- Pearls
 - Unstable VT or VT in the setting of underlying disease such as hypertrophic cardiomyopathy may require an ICD to prevent sudden death

Ectopic Atrial Tachycardia (EAT)

- Definition
 - Regular narrow QRS complex tachycardia
 - Fast atrial rates for age with abnormal P wave morphology

- Variable R-R intervals possible from variable AV conduction
 - Rate response to catecholamines
 - Often gradual onset and cool down
- Additional monitor findings
 - Decreased area under curve of arterial line tracing with hypotension due to rate and possible loss of AV synchrony
- Acute treatment
 - Reduce catecholamine burden (treat fevers, acidosis, pain/agitation)
 - Replete electrolytes
 - Digoxin, esmolol, amiodarone, or procainamide
- Chronic treatment
 - Digoxin, oral beta-blocker, flecainide or amiodarone
- Pearls
 - EAT can be difficult to differentiate from reentry/paroxysmal SVT. Obtain 12 lead ECG to make diagnosis
 - Adenosine, rapid atrial pacing, and cardioversion are not effective in terminating arrhythmia

Multifocal or Chaotic Atrial Tachycardia (MAT)

- Definition
 - Irregular tachycardia with narrow and/ or wide QRS complexes secondary to aberration
 - At least three separate non-sinus P wave morphologies
 - Variable P-R, P-P, and R-R intervals
- Additional monitor findings
 - Variable pulse pressure and degrees of AV dyssynchrony
- Acute treatment
 - Beta-blocker, amiodarone or procainamide continuous infusion
- Chronic treatment
 - Typically a combination of amiodarone, flecainide, digoxin, or beta-blocker
- Pearls
 - Not amenable to overdrive pacing, cardioversion, or adenosine (which may unmask the diagnosis)

Atrial Fibrillation

- Definition
 - Irregularly irregular rhythm with narrow QRS
 - No discernible P waves and variable AV conduction
 - Uncommon in children
- Additional monitor findings
 - CVP: loss of a-wave
- Acute treatment
 - Rate control: beta-blocker (i.e., esmolol), calcium channel blocker (i.e., diltiazem) if over 1 year, or digoxin
 - Rhythm control: cardioversion (0.5-2 J/Kg)

- Chronic treatment
 - Rate control: beta-blocker (i.e. esmolol), calcium channel blocker (i.e. diltiazem) if over 1, or digoxin
 - Rhythm control: amiodarone, sodium channel blocker, or sotalol
- Pearls
 - Obtain TEE prior to cardioversion if you suspect atrial fibrillation for >48 hours to rule out intracardiac thrombus

Bradycardia

- 2nd degree AV block Mobitz I (Wenckebach)
 - Definition
 - Gradually increasing P-R interval with eventual blocked conduction to the ventricle
 - Look for variable P-R and R-R intervals
 - Additional monitor findings
 - Intermittent absent AV conduction
 - Acute treatment: none
 - Chronic treatment: none
 - Pearls
 - Sign of increased vagal tone
 - May be seen during rapid atrial pacing
- 2nd degree AV Block Mobitz II
 - Definition
 - Regularly repeating pattern of normal sinus beats with dropped beats
 - A fixed ratio (2:1, 3:1, 4:1, etc) leads to a constant P-R interval
 - Additional monitor findings
 - Intermittent cannon A waves
 - Acute treatment
 - Treat reversible causes such as ischemia, medications that may impair AV conduction, or vagal tone
 - If asymptomatic, no acute treatment need
 - If symptomatic (i.e. long pauses) then pacing, atropine, or isoproterenol may be considered
 - Chronic treatment
 - Acquired Mobitz II is an indication for permanent pacing
 - Pearls
 - A more advanced grade AV block than Type I raising concern for progression to complete AV block
- Complete heart block
 - Definition
 - Complete absence of AV conduction
 - Atrial rate > ventricular rate
 - Look for variable P-R but regular R-R intervals
 - May be congenital, acquired, or post-surgical in etiology

- Additional monitor findings
 - Absent AV synchrony
- Acute treatment: if symptomatic or hemodynamically significant
 - Isoproterenol
 - Temporary ventricular pacing
- Chronic treatment
 - Post-surgical without expected recovery of AV conduction or persisting at least 7 days, symptomatic bradycardia, prolonged pauses, bradycardia with wide escape rhythm, complex ventricular ectopy, or cardiac dysfunction may require permanent pacemaker
- Pearls
 - Acquired AV block (i.e. Lyme Disease) with likelihood of recovery of conduction generally does not require permanent pacing

Pearls and Pitfalls of Pacing in the ICU

- Indications for short term pacing
 - Bradycardia: AV block, sinus node dysfunction
 - Tachycardia: pace to prevent atrial or ventricular arrhythmias or rapid atrial pace to treat re-entrant arrhythmias
 - Improve hemodynamics by restoring AV synchrony (i.e. JET)
- Methods
 - Transcutaneous pacing
 - Ventricular pacing
 - Painful, can result in skin burn
 - Transesophageal pacing
 - Atrial pacing
 - Use if intact AV conduction
 - Avoid >24 hours to prevent esophageal injury
 - Transvenous
 - Ventricular pacing
 - Temporary epicardial wires
- Definitions
 - Modes
 - First letter: paced chamber (D= Dual, V=Ventricle, A=Atrium, O= neither)
 - Second letter: sensed chamber (D= Dual, V=Ventricle, A=Atrium, O= neither)
 - Third letter: mode (I=Inhibit, D= Dual(triggers and inhibits))
 - Capture threshold: lowest energy (mA) that paces the heart
 - Sensitivity threshold: highest amplitude (mV) that intrinsic heart activity is seen
- Pearls
 - Goal for temporary pacing is hemodynamic stability
 - Consider wire function as sensitivity and capture may worsen over time, test wires daily
 - Oversensing and undersensing result in inappropriate pacing

- If unable to capture and the pace the heart, try reversing the polarity of the pacing leads
- Consider placing additional wires (epicardial or additional skin ground)
- Setting up the pacemaker
- Set sensitivity to at least half the sensitivity threshold – most important for ventricular pacing
- Set output to at least double the capture threshold (minimum of 5 or 10 mA)
- In an emergency, start with a high amplitude (15-20 mA), capture the heart, and stabilize the patient; then assess capture threshold
- If there are changes in lead performance (intermittent or failure to capture), it may not be the device but the patient
- Consider acidosis, effusion and electrolyte derangements which may impact lead functionality
- Pitfalls
 - Temporary atrial wires often don't sense well
 - Atrial pacing is ineffective in the presence of atrial arrhythmias or AV conduction disease
 - Asynchronous ventricular pacing may interfere with intrinsic conduction (R-on-T phenomenon)
 - Rapid atrial pacing NOT to be used in the ventricle

Postoperative Considerations for Patients with Permanent Pacemakers

- Know the manufacturer, type (epicardial vs. transvenous), location of generator (abdominal vs. infraclavicular), indication for device, underlying rhythm, arrhythmia history, cardiac anatomy, pacing mode, rate, and pacing/defibrillation therapy settings
- Inquire about intraoperative complications or concerns that may impact the device functionality

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CHAPTER 4

Pharmacology

Chapter Editors

Ricardo Munoz, MD, FAAP, FCCM, FACC

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Section 1: Vasoactives

Section 2: Antiarrhythmics

Section 3: Sedation and Analgesia

Section 4: Steroid Therapy

Section 5: Diuretics

Section 6: Oral Heart Failure Therapy

- Venodilation and dose dependent systemic and pulmonary arterial vasodilation
- Does not require biotransformation to yield nitric oxide therefore tachyphylaxis does not develop
- Pharmacokinetics
 - Decomposes nonenzymatically to yield cyanide, $t_{1/2} < 5$ minutes
 - Cyanide undergoes transulfuration to form thiocyanate
 - Thiocyanate is renally excreted and has a $t_{1/2}$ of 3 days
 - Thiocyanate accumulation may lead to seizures
 - Adequate thiosulfate (sulfur donor) is needed to prevent cyanide accumulation
- Clinical indications
 - Systolic heart failure
 - Systemic arterial hypertension
- Dosing
 - 0.5-5 mcg/kg/min

Nitroglycerin

- Mechanism of action
 - Undergoes biotransformation to yield NO therefore tachyphylaxis develops
 - Nitric oxide mediates increases in cGMP in vascular smooth muscle cells
 - Dose dependent coronary artery vasodilation
 - Low doses (0.5-3 mcg/kg/min)
 - Venodilation
 - High doses (3-5 mcg/kg/minute)
 - Venodilation, systemic and pulmonary arterial vasodilation
- Mechanism of action
 - Hydrolyzed by hepatic enzymes, $t_{1/2} < 5$ minutes
- Clinical indications
 - Alleviating pulmonary congestion due to CHF
 - In higher doses, increases stroke volume and cardiac output
- Dosing
 - 0.5-5 mcg/kg/min

Arginine Vasopressin

- Mechanism of action
 - Interacts with G protein receptors on vascular smooth muscle cells
 - Physiologic levels needed for normal vascular tone
 - Venoconstriction and dose dependent systemic arterial vasoconstriction
- Pharmacokinetics
 - Metabolized by vasopressinases, $t_{1/2}$ 15 minutes
- Clinical Indications
 - Vasoplegia
- Dosing
 - 0.0002-0.002 units/kg/min

Section 2

Antiarrhythmics

Juliette Hunt, MD, Melanie Muller, MSN, CRNP-AC and
Casey Naughton, MS, CPNP-AC

Procainamide

- Mechanism of action
 - Class IA: Na^+ channel blocker; conduction velocity and automaticity are decreased
- Pharmacokinetics
 - IV: rapid initial distribution phase that is completed in 30 minutes
 - Active metabolite NAPA is renally excreted
 - Therapeutic levels: draw 6-12 hours after infusion initiation
 - Procainamide 4-10 mcg/mL
 - NAPA 15-25 mcg/mL
- Clinical indications
 - Reentry supraventricular and ventricular tachycardias and junctional ectopic tachycardia
- Dosing
 - IV load is 5 to 10 mg/kg over 5 minutes (not to exceed 1mg/kg/min), may repeat every 5 to 10 minutes to maximum total loading dose of 15 mg/kg; do not exceed 500 mg in 30 minutes
 - Continuous IV infusion: 20 to 80 mcg/kg/min; maximum dose: 2 g/day
- Side effects
 - Prolonged QT, torsade de pointes, sinus bradycardia, AV block, and negative inotropy

Lidocaine

- Mechanism of action
 - Class IB: Na^+ channel blocker
 - Slows ventricular conduction but has minimal effect on nodal and atrial tissue
 - Slows the fast diastolic depolarization (phase 0) and shortens the duration of the action potential by accelerating the repolarization
- Pharmacokinetics
 - Rapid onset and short duration, given IV because of hepatic first pass metabolism
- Clinical indication
 - Ventricular tachycardia
- Dosing
 - Bolus: 0.5-1 mg/kg IV/IO/ET, not to exceed 100 mg
 - Continuous IV infusion of 20-50 mcg/kg/min
- Side effects
 - CNS toxicity as levels rise above 10-15 mcg/mL

Flecainide

- Mechanism of action
 - Class IC: Na⁺ channel blocker
 - Marked prolongation of conduction velocity but does not significantly affect the action potential duration
- Pharmacokinetics
 - PO only form available in US with t_{1/2} 8-12 hours
 - Dosed Q 8-12 hours
 - Mostly renally cleared but also metabolized via the hepatic P450 system
 - Therapeutic levels: 0.2-1 mcg/mL
- Clinical indication
 - Reentrant supraventricular tachycardia and ventricular tachycardia
- Dosing
 - Start: 50-100 mg/m²/day, increase dose Q4 days (max: 200 mg/m²/day)
 - Alternate max dosing 3-6 mg/kg/day, divided Q8-12 hours
- Side effects
 - Proarrhythmic; caution use in patients with structural heart disease

Metoprolol

- Mechanism of action
 - Class II: beta blocker, selective inhibitor of beta₁-adrenergic receptors at lower doses
 - Prolongs PR and QT intervals and has little effect on QRS duration
 - Increases refractory period of the AV node
- Pharmacokinetics
 - PO onset of action within 1 hour
 - Extensive metabolism in liver via P450, oral bioavailability is 50%
- Clinical indications
 - Supraventricular and ventricular tachycardia, long QT syndrome
- Dosing
 - 1-2 mg/kg/day PO divided Q12 hours, maximum of 6 mg/kg/day (<200 mg/day)
- Side effects
 - Bradycardia, worsening ventricular function, hypotension, worsening of AV conduction disturbances, bronchospasm, hypoglycemia

Propranolol

- Mechanism of action
 - Class II: beta-adrenergic (beta₁ and ₂ blocker)
- Pharmacokinetics
 - Immediate release PO onset is 1-2 hours
 - Duration 6 hours, t_{1/2} elimination 4-6 hours
 - Metabolism in liver, significant first-pass, 30-40% bioavailability; metabolites primarily excreted in urine

- Clinical indication
 - Supraventricular and ventricular tachycardia
- Dosing
 - Neonates initial PO dose 0.25mg/kg/dose Q 6-8 hours (maximum 5mg/kg/day)
 - Neonates initial IV dose 0.01mg/kg slow IV push over 10 minutes, may repeat every 6-8 hours and increase slowly to a maximum of 0.15mg/kg/dose Q 6-8 hours
 - Children initial PO dose 0.5-1 mg/kg/day in divided doses Q 6-8 hours, titrate upwards every 3-4 days; usual dose is 2-4mg/kg/day (16mg/kg/day or 60mg/day)
 - Children initial IV dose: 0.01-0.1 mg/kg slow IV push over 10 minutes (max 1 mg in infants and 3mg in children)
- Side effects
 - Bradycardia, worsening ventricular function, hypotension, worsening of AV conduction, bronchospasm, hypoglycemia

Esmolol

- Mechanism of action
 - Class II: Selective beta-₁ adrenergic blocker
- Pharmacokinetics
 - Rapid onset and short duration of action with $t_{1/2}$ in minutes
- Clinical indication
 - Reentrant supraventricular tachycardia and ventricular tachycardia
- Dosing
 - 25-500 mcg/kg bolus, then 100-1000 mcg/kg/min
- Side effects
 - Bradycardia, worsening ventricular function, hypotension, worsening of AV conduction, bronchospasm, hypoglycemia

Amiodarone

- Mechanism of Action
 - Class III: inhibits adrenergic stimulation, possesses alpha and beta-adrenergic blocking properties
 - Affects sodium, potassium and calcium channels
 - Prolongs the action potential and refractory period in myocardial tissue
 - Decreases AV conduction and sinus node function
 - Possesses vasodilatory and negative inotropic effects
- Pharmacokinetics
 - Metabolized in the liver via P450 enzymes and possibly the GI tract
- Clinical indication
 - Supraventricular and ventricular tachyarrhythmias and ventricular fibrillation
- Dosing
 - Loading dose: 1-5mg/kg over 25-60 minutes, may repeat
 - Maintenance infusion: 5-20 mg/kg/day

- Side effects
 - Rapid administration of the loading dose may precipitate circulatory collapse
 - Acute: hypotension, torsade de pointes, bradycardia, AV block
 - Long term use may cause thyroid abnormalities and/or optic neuropathy

Verapamil

- Mechanism of Action
 - Class IV: calcium channel blocker
 - Inhibits calcium ions from entering the voltage dependent channels of the vascular smooth muscle and myocardial cell during depolarization
- Pharmacokinetics
 - Metabolized in the liver, $t_{1/2}$ in infants is 4.4-6.9 hours
- Clinical indication
 - Treatment and prevention of supraventricular tachyarrhythmias
- Dosing
 - IV 1-16 years: 0.1-0.3mg/kg/dose to be given over 2-3 minutes, may repeat dose in 30 minutes (maximum first dose: 5mg, maximum second dose: 10mg)
 - Not recommended for children <1 year

Adenosine

- Mechanism of action
 - Class V antiarrhythmic: slows conduction time through the atrioventricular node, interrupting the re-entry pathways through the AV node and restoring normal sinus rhythm
- Pharmacokinetics
 - Metabolized intracellularly with a $t_{1/2}$ of <10 seconds
- Clinical indication
 - Reentrant supraventricular tachycardia involving the AV node
- Dosing
 - IV: initial dose 0.1mg/kg (maximum 6mg/dose), if not effective increase dose by 0.1mg/kg up to 0.3-0.5mg/kg (maximum 12mg/dose)
 - Must be given by rapid IV push
- Side effects
 - Hypotension, sinus arrest, bronchospasm
 - Antagonized by methylxanthines

Digoxin

- Mechanism of action
 - Class V antiarrhythmic: primary mechanism is inhibition of membrane-bound Na^+K^+ ATPase resulting in intracellular calcium loading and its primary cardiac effect is AV conduction delay
- Pharmacokinetics
 - Onset of action: PO 30 minutes-2 hours, IV 5-30 minutes

- Half-life and elimination is dependent upon age, renal, and cardiac function
- Level monitoring not routinely indicated (only for assessing toxicity)
- Clinical indication
 - Treatment and prevention of reentrant SVT
- Dosing
 - See table below for Total Digitalizing Dose (TDD) and maintenance doses in mcg/kg/24 hours
 - Initial: $\frac{1}{2}$ TDD, then $\frac{1}{4}$ TDD Q8-18 hours x 2 doses; obtain ECG 6 hours after dose to assess for toxicity
 - Maintenance: <10 years: give maintenance dose divided BID; >10 years: give maintenance dose QD
 - Please note dosing should be adjusted with renal dysfunction
- Side effects
 - Bradycardia, AV node block, ventricular arrhythmias

Digoxin Dosing (In mcg/kg/day)				
Age	Total digitalizing dose (TDD)		Daily maintenance	
Age	PO	IV/IM	PO	IV/IM
Premature Neonate	20	15	5	3-4
Full-term Neonate	30	20	8-10	6-8
1 mo - <2 yr	40-50	30-40	10-12	7.5-9
2 - 10 yr	30-40	20-30	8-10	6-8
>10 yr and <100 kg	10-15	8-12	2.5-5	2-3

Section 3

Sedation and Analgesia

Joanna Rosing, MD, Matt Bochkoris, MD, Erica Lin, MD, FAAP, Anna Brown, MD and Jamie Schwartz, MD, FAAP

Midazolam

- Mechanism of action
 - Binds to GABA α -receptor, facilitating the inhibitory effects of the GABA molecule
- Pharmacokinetics
 - Rapid onset of action, short elimination $t_{1/2}$
 - Hepatic metabolism, accumulation of active metabolites with repeated administration may occur, some of which are renally excreted
- Cardiopulmonary effects
 - Respiratory: respiratory depressant
 - Cardiovascular: by virtue of providing anxiolysis and sedation, the release of endogenous catecholamines decreases which may be of hemodynamic significance
- Clinical indication
 - Anxiolysis, sedative-hypnotic, anticonvulsant, antegrade amnestic
- Dosing
 - Intermittent bolus: 0.05-0.1 mg/kg
 - Duration of action approximately 1 hour
 - Infusion, initial: 0.05-0.1 mg/kg/hour
 - May require higher dosing as tolerance develops
- General considerations
 - Tolerance for and dependence on benzodiazepines can develop

Lorazepam

- Mechanism of action
 - Binds to GABA α -receptor, facilitating the inhibitory effects of the GABA molecule
- Pharmacokinetics
 - Relative to midazolam, slower onset of action and relatively longer $t_{1/2}$
 - Undergoes hepatic metabolism producing inactive metabolites
- Cardiopulmonary effects
 - Respiratory: respiratory depressant
 - Cardiovascular: by virtue of providing anxiolysis and sedation, the release of endogenous catecholamines decreases which may be of hemodynamic significance
- Clinical indication
 - Anxiolysis, sedative-hypnotic, anticonvulsant, antegrade amnestic
- Dosing
 - Intermittent bolus: 0.05-0.1 mg/kg
 - Duration of action approximately 2-4 hours
 - Infusion, initial: 0.05-0.1 mg/kg/hour

- May require higher dosing as tolerance develops
- General considerations
 - Tolerance for and dependence on benzodiazepines can develop
 - Prolonged use may lead to the accumulation of the diluent propylene glycol causing metabolic acidosis, renal failure, and CNS depression. Propylene glycol is metabolized in the liver and excreted unchanged in the urine

Fentanyl

- Mechanism of action
 - Synthetic opioid, binds μ -receptors (mu receptors)
- Pharmacokinetics
 - Rapid onset due to high lipid solubility
 - Duration of action short due to redistribution from the CNS
 - With repeated dosing and infusions, significant accumulation within muscle and fat occurs where it is slowly released, producing a much longer elimination $t_{1/2}$
 - Metabolized in the liver producing inactive metabolites
- Cardiopulmonary effects
 - Respiratory: respiratory depressant, risk of chest wall rigidity
 - Cardiovascular
 - Moderate bradycardia
 - Does not cause histamine release
 - By virtue of providing some degree of sedation, endogenous catecholamine release may decrease, which may be of hemodynamic significance
- Clinical indication: analgesia
- Dosing
 - Intermittent bolus: 0.5-1 mcg/kg
 - Duration of action approximately 1 hour
 - Infusion, initial: 0.5-1 mcg/kg/hour
 - May require higher dosing as tolerance develops
- General considerations
 - Tolerance for and dependence on opioids may develop

Morphine

- Mechanism of action
 - Opioid analgesic mainly via supraspinal μ -1 receptor activation
- Pharmacokinetics
 - Poor lipid solubility compared to other opioids
 - Hepatic metabolism with renally excreted active metabolites
- Cardiopulmonary effects
 - Respiratory: respiratory depressant
 - Cardiovascular: may cause histamine release resulting in vasodilation of venous capacitance and arterial resistance vessels

- Clinical indication: analgesia
- Dosing
 - Intermittent bolus: 0.05 – 0.1 mg/kg
 - Duration of action approximately 2 hours
 - Infusions, initial: 0.05-0.10 mg/kg/hour
 - May require higher dosing as tolerance develops
- General considerations
 - Tolerance for and dependence on opioids may develop
 - Additional side effects include: constipation, urinary retention, and nausea

Methadone

- Mechanism of action
 - Synthetic opioid
- Pharmacokinetics
 - High lipid solubility with a large volume of distribution (V_D) and long $t_{1/2}$ (19 hours-60 hours)
- Clinical indication: weaning protocol for treating opioid dependence
- Dosing
 - 0.05-0.2 mg/kg IV/ PO Q12-24 hours with the initial range and duration of wean determined by opioid exposure history

Dexmedetomidine

- Mechanism of action
 - α_2 adrenergic agonist in the CNS producing sedation, and anxiolysis, and central sympatholysis
 - Weak analgesic effects primarily mediated through α_2 stimulation in the spinal cord
- Pharmacokinetics
 - Elimination $t_{1/2}$ of 2 hours
 - Metabolized by liver producing inactive metabolites
 - Prolonged clearance in neonates
- Cardiopulmonary effects
 - Respiratory: minimal respiratory depression
 - Cardiovascular: central sympatholysis induced bradycardia and decrease in systemic vascular resistance
- Clinical indication: sedation, mild analgesic properties
- Dosing
 - Loading dose 0.5-1 mcg/kg
 - Infusion: 0.2-1 mcg/kg/hour
 - Intermittent bolus: 0.2-1 mcg/kg
- General considerations
 - Unclear whether withdrawal phenomena occur

Propofol

- Mechanism of action
 - Binds to the GABA β -subunit enhancing inhibitory neurotransmission
 - Capable of providing deep sedation and burst suppression
- Pharmacokinetics
 - Highly lipid soluble agent with rapid onset of action
 - Duration of activity short due to redistribution from the CNS
 - Metabolized in the liver with renal excretion
- Cardiopulmonary effects
 - Respiratory: respiratory depressant
 - Cardiovascular: suppression of endogenous catecholamine release
- Clinical indication: general anesthetic, sedation
- Dosing
 - Induction of anesthesia: 1-3 mg/kg IV
 - Infusion: 25-100 mcg/kg/minute
- General considerations
 - Hyperlipidemia and hypertriglyceridemia may occur due its delivery in a lipid emulsion
 - Propofol infusion syndrome: characterized by a refractory metabolic acidosis, typically seen with high doses or use >48 hours

Ketorolac

- Mechanism of action
 - Inhibition of prostaglandin synthesis, attenuation of inflammation
- Pharmacokinetics
 - Hepatic metabolism
- Cardiopulmonary effects: none
- Clinical indication: Non-steroidal anti-inflammatory analgesic and antipyretic
- Dosing
 - Children 2-16 years or <50 kg: 0.5 mg/kg IV (max 15 mg/dose) Q 6-8 hours
 - > 16 years or >50kg: 30mg IV Q6-8 hours (max 120 mg/day)
- General considerations
 - Use with caution in patients who have (or are at risk for) renal insufficiency
 - Causes platelet inhibition, use with caution in bleeding patients
 - Use with caution in neonates
 - Not recommended for use >5 days

Ketamine

- Mechanism of action
 - Antagonism of NMDA receptors and agonism at opioid receptors
- Pharmacokinetics
 - Rapid onset of action
 - Hepatic metabolism with excretion of metabolites into urine
 - Metabolized to norketamine (active metabolite)

- Cardiopulmonary effects
 - Respiratory: no respiratory depression
 - Cardiovascular: stimulates endogenous catecholamine release, cardiomyocyte depressant
- Clinical indication: intravenous and intramuscular analgesic, sedative, and anesthetic
- Dosing
 - Induction: 1-2 mg/kg IV, 4-8 mg/kg IM
- General considerations
 - Emergence phenomena or hallucinations: particularly in older children, premedication with benzodiazepines may significantly reduce the severity of such episodes

Section 4

Steroid Therapy

Juliette Hunt, MD and Aimee Shumard, CCRN, MSN, CPNP-AC

Steroids (Glucocorticoids)

- Mechanism of action
 - Glucocorticoids are synthetic steroids that mimic the action of cortisol
 - Glucocorticoids bind to and activate receptors within the cytoplasm followed by migration into the nucleus where they alter DNA transcription and protein synthesis
 - Glucocorticoid signaling also occurs through membrane-associated receptors and 2nd messenger pathways (non-genomic mechanism)
 - Anti-inflammatory effect
 - Required for cardiovascular homeostasis
 - Vascular tone (primary effect)
 - May enhance adrenergic receptor response to catecholamines
 - May have some effect on cardiomyocyte function
- Clinical indication
 - Preoperative
 - Used to ameliorate the inflammatory response to cardiopulmonary bypass during cardiac surgery
 - Data on postoperative outcomes are conflicting
 - Postoperative
 - May be used for refractory hypotension due to an inadequate adrenal response
 - Prevention or treatment of post-extubation croup (anti-inflammatory effect)
- Dosing
 - Hydrocortisone
 - Stress dose: 50mg/m²/day IV divided Q6 hours
 - Shock dose: initial 1-2 mg/kg/day IV divided Q 6 hours, may titrate up to 50mg/kg/day
 - Dexamethasone
 - Post-extubation croup: 0.5mg/kg IV Q6 hours for 48-72 hours

Section 5

Diuretics

David Kwiatkowski, MD, Meaghan Doherty, MD and
Alejandro Lopez-Magallon, MD

Loop Diuretics: Furosemide, Bumetanide

- Mechanism of action: actively transported into the lumen to block $\text{Na}^+/\text{K}^+/\text{Cl}^-$ co-transporter in ascending loop of Henle
- Side effects: hypokalemia, hyponatremia, hypocalcemia, metabolic alkalosis, ototoxicity
- General considerations
 - Most potent class of diuretics
 - Ethacrynic acid infrequently used due to high incidence of ototoxicity
 - Bioavailability of furosemide is highly variable and oral doses may not have similar efficacy as IV doses

Loop Diuretic Dosing				
Medication	Age	Oral	IV/IM	Continuous Infusion
Furosemide (Lasix)	Neonates and preterm newborns	0.5-2 mg/kg/dose Qday to QID	0.5-2 mg/kg/dose Qday to QID	
	Term newborn, infants and children	0.5-2 mg/kg/dose Qday to QID	0.5-2 mg/kg/dose Qday to QID	0.05 to 0.5 mg/kg/hour
	Adults	20-80mg/dose	20-40mg/dose, may increase by 20mg and repeat in 1-2 hours as needed	0.05to 0.5 mg/kg/hour
Bumetanide (Bumex)	Infants and children	0.01-0.1 mg/kg/dose Qday to BID max: 10mg/day	0.01-0.1mg/kg/dose Qday to BID max: 10mg/day	Total daily IV dose can be given as continuous infusion over 24 hours
	Adults	0.5-1mg/dose, Qday to BID max: 10mg/day	0.5-1mg/dose max: 10mg/day	0.9-1mg/hour

Thiazide Diuretics: Chlorothiazide, Hydrochlorothiazide, Metolazone

- Mechanism of Action
 - Blocks NaCl co-transporter in the distal convoluted tubule

- May be used synergistically with loop diuretics
- Side effects: hypokalemia, hypercalcemia, hypochloremic alkalosis, interstitial nephritis
- General considerations: hydrochlorothiazide is only available orally, chlorothiazide is available orally and intravenously

Thiazide Diuretic Dosing			
Medication	Age	Daily Dose	Maximum Daily Dose
Chlorothiazide (Diuril)	Infants and children	PO: 10-30 mg/kg/day divided Qday to BID IV: 2-10mg/kg/day divided Qday to BID	IV: 20mg/kg
	Adults	Oral: 250-500mg/dose Qday to BID IV: 100-1000mg/dose Qday to BID	PO and IV: 2000mg
Hydrochlorothiazide (Hydrodiuril)	<6 months	PO: 1-3 mg/kg/day divided Qday to BID	37.5mg
	> 6 months	PO: 1- 2mg/kg/day divided Qday to BID	6 months-2 years: 37.5mg 2-12 years: 100mg
	Adults	PO: 25-100mg/day divided Qday to BID	200mg

Metolazone: Zaroxolyn

- Mechanism of action: inhibits sodium resorption in the distal tubules similar to thiazide diuretics
- Dosing
 - Children: 0.2-0.4 mg/kg/day PO divided Qday to BID
 - Adults: 2.5-10mg/dose PO Qday to BID, maximum dose 20mg/day
- Side effects: hyponatremia, hypokalemia, hypercalcemia, hypochloremic alkalosis. Caution use in patients with sulfonamide allergies

Spirolactone: Aldactone

- Mechanism of action: aldosterone antagonist
 - Decreases expression of Na⁺ channels and Na⁺/K⁺ ATPase in collecting tubules
 - Weak diuretic, used primarily as a potassium sparing agent
- Dosing
 - Neonates: 1-3 mg/kg/day PO divided Qday to BID
 - Children: 1.5-3.5 mg/kg/day PO divided Qday-QID
 - Adults: 25-200 mg/day PO divided Qday to BID
- Side effects: hyperkalemia, caution use in conjunction with ACE inhibitors or other medications that inhibit potassium excretion

Acetazolamide: Diamox

- Mechanism of action: carbonic anhydrase inhibitor
 - Inhibits breakdown of NaHCO_3 in the proximal collecting tubule
 - Weak diuretic
 - May be used to treat metabolic alkalosis
- Dosing
 - Children: 5mg/kg/dose PO or IV Qday to QID
 - Adults: 250-375mg/dose PO or IV Qday or QID
- Side effects: metabolic acidosis, hyponatremia, hypokalemia, caution use in patients with sulfonamide allergies

Section 6

Oral Heart Failure Therapy

Thomas D. Ryan, MD, PhD, BreAnn Taylor, PharmD, BCPS and Ivan Wilmot, MD

Angiotensin Converting Enzyme (ACE) Inhibitors: Captopril, Enalapril, Lisinopril

- Mechanism of action
 - Competitive inhibitor of angiotensin converting enzyme, preventing conversion of angiotensin I to angiotensin II
 - Mitigate angiotensin II-mediated vasoconstriction and fluid retention
- Clinical indication
 - Hypertension
 - Afterload reduction in patients with systolic dysfunction and failure
 - Promotes ventricular remodeling in patients with systolic dysfunction
- Dosing
 - Captopril: 0.3-6 mg/kg/day PO divided Q8 hours, maximum 150 mg/day
 - Enalapril: 0.1-0.5 mg/kg/day PO divided BID
 - Lisinopril: 0.07-0.5 mg/kg/dose PO Qday, maximum dose 40 mg/day

Angiotensin Receptor Blockers (ARB): Losartan

- Mechanism of action
 - ARB blockade: selective and competitive angiotensin II receptor antagonist
 - Mitigate angiotensin II-mediated vasoconstriction and fluid retention
- Clinical indication
 - Hypertension
 - Afterload reduction in patients with systolic dysfunction and failure
 - Promotes ventricular remodeling in patients with systolic dysfunction
- Dosing
 - Losartan
 - 6-16 years of age: 0.7-1.4 mg/kg/dose PO Qday, maximum 100 mg/day
 - >17 years: 25-100 mg/day PO divided Qday to BID

Beta blockers: Propranolol, Atenolol, Metoprolol, Carvedilol

- Mechanism of action
 - Propranolol, carvedilol: non-selective β -adrenergic blocker (carvedilol also has α -adrenergic blocking activity)
 - Atenolol, metoprolol: selective inhibitor of cardiac specific β_1 -adrenergic receptors
 - Negative inotrope and chronotrope
- Clinical indication
 - Promotes ventricular remodeling in patients with systolic dysfunction and failure
 - Slows heart rate and increases diastolic time in patients with ventricular hypertrophy and impaired diastolic filling

- Dosing
 - Propranolol
 - Infants: 0.25-1 mg/kg/dose PO Q6-8 hours, max daily dose 5mg/kg
 - Children: 0.5-4 mg/kg/day PO divided Q 8-12 hours, maximum 16 mg/kg/day or 60 mg/day
 - Adults: 80-240 mg/day PO divided BID to TID, maximum 640 mg/day
 - Atenolol
 - Children: 0.5-2 mg/kg/day PO divided Qday to BID
 - Adults: 25-100 mg PO Qday
 - Metoprolol
 - Children (immediate release, metoprolol tartrate): 1-2 mg/kg/day PO divided BID, increase to maximum of 6 mg/kg/day or 200 mg/day
 - Children (extended release, metoprolol succinate): 25-100 mg PO Qday
 - Carvedilol
 - Children: 0.05-0.2 mg/kg/day PO divided BID, titrate every 2 weeks to 0.4-0.8 mg/kg/day, maximum 50 mg/day
 - Adults: 3.125 mg PO BID for 2 weeks, titrate to maximum 50 mg PO BID

Mineralocorticoid/Aldosterone Receptor Antagonists: Spironolactone

- Mechanism of action: competes with aldosterone in distal renal tubule
- Clinical indication
 - Weak diuretic
 - Primarily used for potassium-sparing effects
- Dosing
 - <25kg: 1-3 mg/kg/day PO divided BID
 - ≥ 25 kg: 25 mg PO Qday

Calcium Channel Blockers: Amlodipine, Diltiazem, Verapamil

- Mechanism of Action
 - Prevent calcium ion entry into vascular smooth muscle cell and cardiomyocyte, producing relaxation of smooth muscle and coronary vasculature
- Clinical indication
 - Hypertension
 - Afterload reduction in systolic dysfunction and failure
- Dosing
 - Amlodipine: 0.1-1 mg/kg/day PO divided in Qday to BID, adult max: 10mg daily
 - Diltiazem (children): 1.5-3.5 mg/kg/day PO divided TID to QID (
 - Verapamil
 - 1-5 years: 4-8 mg/kg/day PO divided TID
 - > 5years: 80 mg PO TID

Digoxin

- Mechanism of action
 - Inhibits Na/K ATPase resulting in increased intracellular calcium and increased contractility
- Clinical indication
 - Consider in individuals with symptomatic heart failure and low ejection fraction currently receiving ACE/ARB and beta blocker
 - Arrhythmia control (see Chapter 4 Section 2: Antiarrhythmics)
- Dosing
 - Variable by age: 2.5-15 mcg/kg/day
 - In general no loading dose for heart failure
 - See Chapter 4 Section 2: Antiarrhythmics for dosing for arrhythmia control



CHAPTER 5

Acyanotic Lesions with Increased Pulmonary Blood Flow

Chapter Editors

Aarti Bavare, MD, MPH, FAAP

Amy Donnellan, APRN

Section 1: Patent Ductus Arteriosus

Section 2: Aortopulmonary Window

Section 3: Atrial Septal Defect

Section 4: Ventricular Septal Defect

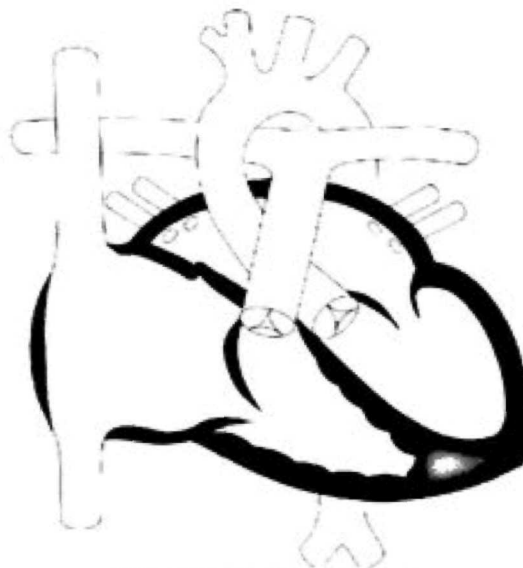
Section 5: Atrioventricular Canal Defect

Section 6: Truncus Arteriosus

Section 1
Patent Ductus Arteriosus
Elizabeth Price, PA-C

Anatomy

- Ductus arteriosus is a communication between the pulmonary artery and the aorta
- In utero, the ductus arteriosus shunts blood from the pulmonary artery to the aorta, away from the lungs
- After birth, the increase in oxygen saturation and decrease in circulating prostaglandin causes functional closure of the ductus arteriosus
- Failure of the ductus arteriosus to close following birth is called patent ductus arteriosus (PDA)



Patent Ductus Arteriosus

Preoperative Physiology

- Left to right shunt through the PDA leads to increased pulmonary blood flow, LA dilation and volume load to the LV
- The degree of left to right shunt through the PDA is determined by the resistance of the duct (diameter, length, and tortuosity) and difference between systemic vascular resistance and pulmonary vascular resistance
- A moderate to large left to right shunt will present with signs of congestive heart failure including: failure to thrive, increase work of breathing, and cardiomegaly
- Pulmonary hypertension may develop in the first year of life if untreated
- Degree of clinical heart failure dictates need for medical management of CHF symptoms including: diuretics and fortified feeds to promote growth

Operative or Interventional Procedures

- Catheter coil/device closure performed in the cardiac catheterization laboratory
- Surgical ligation or division remains the most common method used for PDA closure in newborns
- Performed through a left posterolateral thoracotomy without cardiopulmonary bypass
- Video-assisted thoracoscopic clip ligation can be performed for a ductus with an adequate length

Postoperative Considerations and Management

- Decreased LV volume load postoperatively, improved cardiac output
- Expect reasonable hemodynamics, given no cardiopulmonary bypass
- Treat pulmonary edema from preoperative shunt with diuretics
- Manage pain with intermittent dosing of acetaminophen and opioids

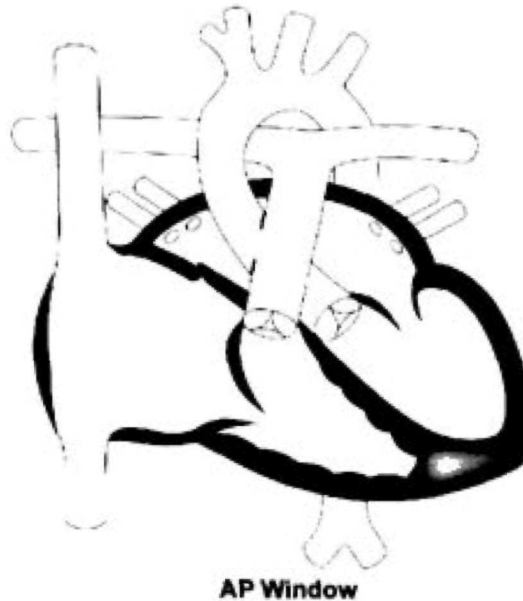
Lesion Specific Postoperative Concerns

- Post catheterization concerns
 - PA coil embolization
 - Hemolysis
 - Left PA stenosis
 - Femoral vessel occlusion
- Postoperative concerns
 - Unintended ligation of major structures: aorta, left pulmonary artery, left main stem bronchus
 - Recurrent laryngeal nerve injury leading to vocal cord paralysis
 - Pneumothorax, chylothorax and atelectasis

Section 2
Aortopulmonary Window
Elizabeth Price, PA-C

Anatomy

- Failure of the aortopulmonary septum formation, causing a communication between the ascending aorta and the pulmonary artery



Preoperative Physiology

- Left to right shunt through the defects leads to increased pulmonary blood flow, LA dilation and volume load to the LV
- The degree of left to right shunt is determined by the size of the defect and pulmonary vascular resistance
- Heart failure symptoms rapidly worsen as pulmonary vascular resistance falls
- A moderate to large left to right shunt will present with signs of congestive heart failure including: failure to thrive, increase work of breathing, and cardiomegaly
- Pulmonary hypertension may develop if untreated
- Degree of clinical heart failure dictates need for medical management of CHF symptoms including: diuretics and fortified feeds to promote growth

Operative Procedure

- Aortopulmonary window does not close spontaneously, surgical treatment is the mainstay of therapy and should be performed early to prevent irreversible pulmonary hypertension
- Surgical patch closure with cardiopulmonary bypass and aortic cross-clamp

Postoperative Considerations and Management

- Decreased LV volume load postoperatively, improved cardiac output
- Inotropic support and volume resuscitation for low cardiac output syndrome following cardiopulmonary bypass
- Mechanical ventilation and inhaled nitric oxide may be necessary to manage pulmonary hypertension episodes
- Treat pulmonary edema from preoperative shunt with diuretics
- Manage pain with intermittent dosing of acetaminophen and opioids
- May require continuous intravenous sedation and analgesia in the immediate postoperative period

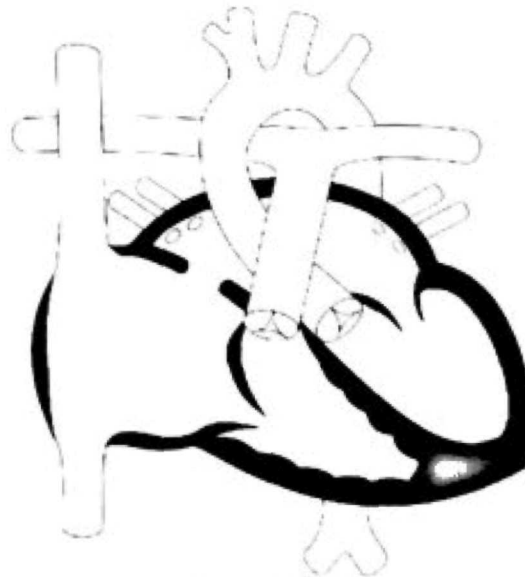
Lesion Specific Postoperative Concerns

- Pulmonary hypertension crisis: manage with 100% FiO₂, iNO, sedation, fluid administration, and phenylephrine
- Distortion of coronary artery, aortic obstruction and pulmonary artery obstruction

Section 3
Atrial Septal Defect
Parag Jain, MD

Anatomy

- Ostium secundum: results from deficiency of septum primum (most common type)
 - Located in the center of the atrial septum.
- Sinus venosus
 - Superior: high in the atrial septum at SVC-RA junction, associated with partially anomalous pulmonary veins (PAPVR)
 - Inferior: low at the IVC/RA junction, associated with Scimitar syndrome
- Ostium primum: located low in the atrial septum, associated with a cleft left atrioventricular valve and other endocardial cushion defects
- Coronary sinus defect: failure of formation of the common wall separating the LA from the coronary sinus
- Patent foramen ovale: inadequate fusion of septum primum and secundum



Atrial Septal Defect

Preoperative Physiology

- Degree and direction of shunt determined by size of defect and compliance of both ventricles, normally results in a left to right shunt
- Usually does not cause significant heart failure symptoms or pulmonary vascular disease until teenage or adult years
- Saturation in the RA higher than SVC or IVC saturation
- RA and RV volume overload

Operative or Interventional Procedures

- Device occlusion in the catheterization lab is an option for small ostium secundum defect
- Repair of ostium primum, ostium secundum, and coronary sinus defect involves either primary stitch closure or patch closure with Dacron or pericardial patch
- Warden procedure indicated for high sinus venosus defect, includes division and reanastomosis of SVC to RA and ASD closure that baffles partial anomalous pulmonary veins to the LA

Postoperative Considerations and Management

- Decreased volume load of ventricles and improved cardiac output
- Usually no need for inotropic support
- Patients usually extubated in operating room or early in postoperative period

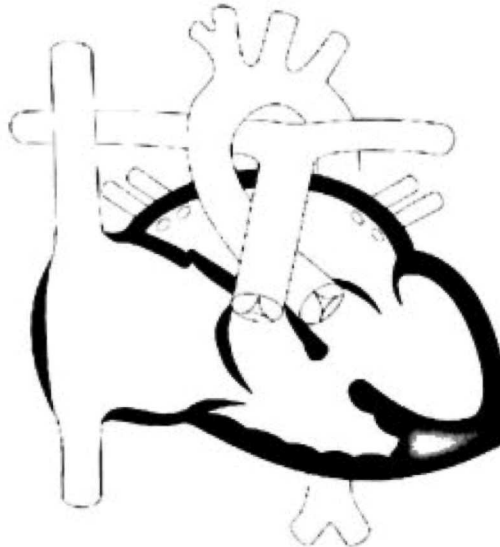
Lesion Specific Postoperative Concerns

- Sinoatrial (SA) node dysfunction, atrial arrhythmias, and junctional rhythm secondary to suturing in the septum and inflammation
- Residual ASD, usually not hemodynamically significant
- Post-pericardiotomy syndrome is characterized by fever, lymphocytosis, nausea, vomiting, abdominal pain, friction rub, and cardiomegaly associated with pericardial effusion. Initial treatment includes aspirin, non-steroidal anti-inflammatory medications (NSAIDs), volume resuscitation, and pericardiocentesis if needed for hemodynamic instability
- LV dysfunction may be noted in older children or adults as a result of chronic RV dilation
- Pulmonary venous obstruction after repair of sinus venosus ASD

Section 4
Ventricular Septal Defect
Sebastian Tume, MD and Krishna Siruguppa, MD

Anatomy

- Perimembranous: subaortic, membranous, or infracristal, located in the LV outflow tract beneath aortic valve
- Outlet: infundibular, supracristal, conal, or subpulmonic, located in the RV outflow tract beneath pulmonary valve, associated with prolapse of aortic valve
- Inlet: atrioventricular canal type, posteriorly located beneath the septal leaflet of tricuspid valve, associated with straddling of AV valve chordae
- Muscular: frequently multiple in the central, apical, or marginal location of the septum



Ventricular Septal Defect

Preoperative Physiology

- Left to right shunt determined by size of the defect and difference between systemic vascular resistance and pulmonary vascular resistance
- Restrictive (defect smaller than aortic root): high resistance to flow, leading to small left to right shunt and normal RV pressures
- Unrestrictive (large defect): no systolic pressure gradient between LV and RV, systemic pressures in both ventricles
- Large left to right shunt leads to increased pulmonary blood flow, LA dilation, LV volume load, and signs of congestive heart failure including: failure to thrive, increase work of breathing and cardiomegaly
- Unrepaired defects with significant pulmonary overcirculation can result in pulmonary vascular obstructive disease and Eisenmenger's syndrome
- Medical management of heart failure symptoms: diuretics, +/- digoxin and fortified feeds to promote growth

- Suspect pulmonary hypertension with a large VSD and no symptoms of heart failure (more common in patients with Trisomy 21 and those repaired at older age)
- Need for cardiac catheterization typically only indicated when echocardiography and clinical findings suggest advanced pulmonary vascular disease

Operative or Interventional Procedures

- Catheter delivered device closure considered for muscular defects and certain perimembranous lesions
- Stitch or Dacron patch closure of the defect, depending on the size

Postoperative Management

- Decreased volume load and increased afterload on the LV
- May need inotropic support and/or afterload reduction for initial 12-24 hours after CPB
- Usually extubated in the operating room or within the first 24 hours postoperatively
- Infants may require a longer period of mechanical ventilation

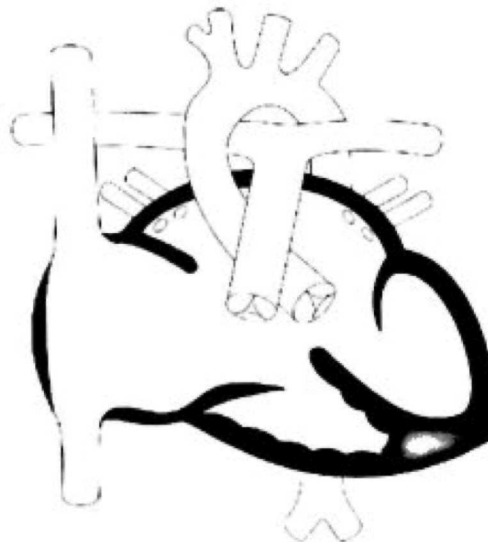
Lesion Specific Postoperative Concerns

- Arrhythmias: junctional ectopic tachycardiac and heart block most common
- Pulmonary hypertension may manifest as low cardiac output or RV dysfunction.
- Residual VSD: may result in inability to diurese and difficulty weaning from mechanical ventilation
- AV valve regurgitation and aortic insufficiency: most likely with subpulmonic VSD and defect with aortic valve prolapse preoperatively

Section 5
Atrioventricular Canal Defect
Ann Scott RN, MSN, CPNP, APNP and
Kimberly Moore RN, MSN, CPNP-AC, CPNP-PC

Anatomy

- Partial: ostium primum ASD, two distinct atrioventricular valves and cleft in left AV valve
- Transitional: ostium primum ASD, restrictive inlet VSD and two atrioventricular valve annuli
- Complete (CAVC): ASD, VSD, common atrioventricular valve
- Both atrioventricular valves attach at same level to ventricular septal crest and the aortic valve is displaced anteriorly resulting in "gooseneck deformity"
- Balanced if equal AV inlet into both ventricles. If unbalanced, one ventricle is usually hypoplastic
- CAVC: Rastelli type determined by attachment of anterior bridging leaflet.
 - Type A: anterior bridging leaflet attaches to ventricular septal crest
 - Type B: anterior bridging leaflet with straddling attachments to the ventricular septum
 - Type C: anterior bridging leaflet is unattached



Complete Atrio-Ventricular Canal Defect

Preoperative Physiology

- Left to right shunt determined by difference between systemic vascular resistance and pulmonary vascular resistance
- Other considerations include: AV valve insufficiency and left ventricular outflow tract obstruction

- Unbalanced CAVC associated with ventricular hypoplasia and single ventricle physiology
- Medical management of heart failure symptoms include: diuretics, +/- digoxin, and fortified feeds to promote growth
- Limited symptoms of heart failure should raise concerns for elevated pulmonary vascular resistance

Operative Procedure

- Surgical intervention is indicated at 3-6 months to avoid progression of pulmonary vascular disease
- Repair consists of ASD and VSD closure, construction of 2 competent AV valves and repair of associated defects
- Techniques
 - Single patch: the ASD and VSD are closed using one patch, bridging leaflets are divided and suspended from the patch and AV valve cleft closed.
 - Two patch: the ASD and VSD are closed using two separate patches, and the AV valve cleft is sutured closed.
 - Australian patch (simplified single patch): the VSD is closed by directly suturing the common AV valve leaflets to the top of the ventricular septum and a patch is used to close the ASD

Postoperative Considerations and Management

- Repair expected to decrease ventricular volume overload and improve hemodynamics
- Inotropic support may be needed for the initial hours after CPB depending on degree of heart failure preoperatively
- Afterload reduction may be needed in the setting of residual AV valve regurgitation
- Patients with preoperative pulmonary hypertension at risk for low cardiac output symptoms if there are no residual defects to allow right to left shunt
 - iNO and sedation beneficial
 - Maintain normal pH and adequate ventilation
- Residual atrial septal defects allow for right to left shunt and maintenance of cardiac output with associated cyanosis

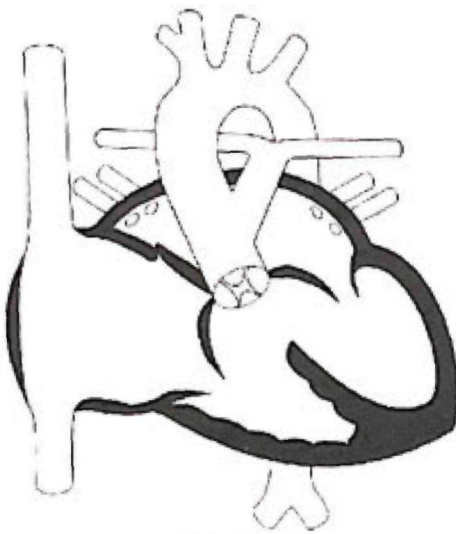
Lesion Specific Postoperative Concerns

- Residual AV valve insufficiency
- Increased LV afterload
- Arrhythmias: specifically heart block and junctional ectopic tachycardia
- Pulmonary hypertension

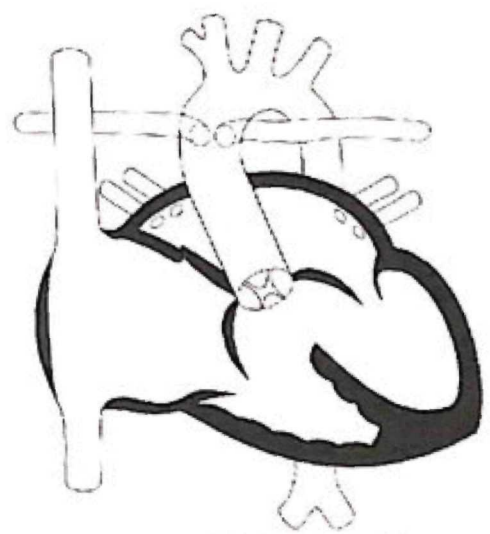
Section 6
Truncus Arteriosus
Parag Jain, MD

Anatomy

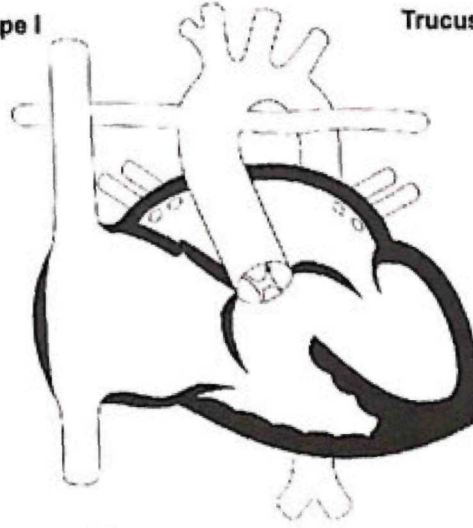
- Single great artery (truncus) arises from the heart
- The truncus receives blood from both ventricles and supplies blood to systemic, pulmonary, and coronary arteries
- Obligate ventricular septal defect
- Types are determined by pulmonary artery origin
 - Type I: single main PA arises from the truncus
 - Type II: separate right and left PA branches arise from the posterior wall of the truncus
 - Type III: separate right and left PA branches arise from the truncus further apart from each other
 - Type IV: associated with interrupted aortic arch, the ductus supplies the descending aorta and the PAs arise from the truncus



Truncus Arteriosus Type I



Truncus Arteriosus Type II



Truncus Arteriosus Type III

Preoperative Physiology

- Systemic and pulmonary venous blood completely mixes
- Arterial oxygen saturation determined by amount of pulmonary blood flow, which is determined by pulmonary vascular resistance
- A large left to right shunt causes increased pulmonary blood flow and volume load of the LA and LV, and eventually causes increased pulmonary vascular resistance
- Develop signs of congestive heart failure once PVR falls after birth

Operative Procedure

- Usually repaired in the neonatal period because of rapid development of congestive heart failure and risk of pulmonary hypertension
- Involves separation of PAs from truncus, establishment of RV to PA continuity, closure of VSD, and repair of associated anomalies

Postoperative Considerations and Management

- RV dysfunction should be expected given the elevated pulmonary pressures preoperatively
- Atrial communication may be left open to allow for a right to left shunt in the immediate postoperative period
- Milrinone, oxygen, and iNO should be considered to support the RV and reduce PVR
- A full repair will decrease the volume load on the LV and increase the afterload on the RV
- Will likely need inotropic support and mechanical ventilation until the chest is closed and/or the hemodynamics improve postoperatively

Lesion Specific Postoperative Concerns

- RV dysfunction and pulmonary hypertension: prevent pulmonary hypertensive crisis by avoiding acidosis, hypothermia, hypoxemia, and agitation
 - Use oxygen, iNO and afterload reduction to support the RV
- Truncal valve dysfunction: use of afterload reduction may be indicated
- Consider residual PA obstruction/distortion or conduit dysfunction in the setting of prolonged RV dysfunction
- Injury to coronary arteries causing LV dysfunction
- Injury to conduction system causing arrhythmias (right bundle branch block, JET, complete heart block, ventricular ectopy)

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CHAPTER 6

Acyanotic Lesions with Normal Pulmonary Blood Flow

Chapter Editors

David Hehir, MD, MS

Lauren Westgate, RN, CPNP-AC/PC, CCRN

Section 1: Aortic Stenosis

Section 2: Coarctation of the Aorta

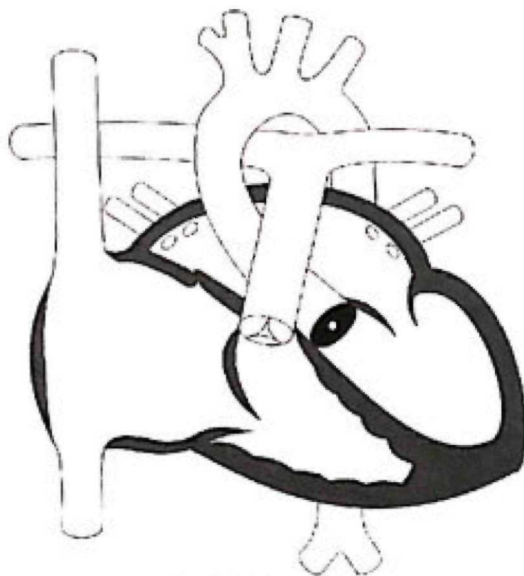
Section 3: Interrupted Aortic Arch

Section 4: Mitral Valve Stenosis

Section 1
Aortic Stenosis
Aimee Jennings RN, CPNP

Anatomy

- Narrowing of aortic valve due to abnormal valve leaflets or small annulus
- Subaortic stenosis associated with normal aortic valve leaflets and annular size with discrete subaortic membrane or tissue
- Supravalvar stenosis is associated with discrete narrowing above sinotubular junction, associated with William's Syndrome
- Congenital causes
 - Neonatal leaflets are thickened, fused and a unicommissural valve is common
 - May be associated with more complex disease (e.g. hypoplastic left heart syndrome, Shone's complex)
- Older children often have bicuspid thickened leaflets with commissural fusion
 - Acquired causes: acute rheumatic fever, endocarditis, connective tissue and inflammatory diseases
- Associated lesions
 - Aortic insufficiency
 - Additional left sided obstructive lesions (mitral stenosis, hypoplastic left ventricle, subaortic stenosis, supravalvar aortic stenosis, aortic arch hypoplasia, coarctation of the aorta)
 - Ventricular septal defect
- Echocardiographic assessment of disease severity defined by mean gradient
 - <30 mmHg = mild
 - 30-50 = moderate
 - >50 = severe
- Critical AS defined by ductal dependent systemic blood flow



Aortic Stenosis

Preoperative Physiology

- Mild to moderate AS well tolerated and may be managed conservatively
 - Mild AS unlikely to require intervention
 - Concentric left ventricular hypertrophy develops over time due to increased afterload
 - Progressive rise in end diastolic pressure and diastolic dysfunction
 - Moderate AS may progress to severe AS and require intervention
- Severe AS/Critical AS requires immediate intervention in the neonate
 - Patent ductus provides systemic output with right to left shunt
 - Left ventricle is usually dilated with very depressed contractility with signs of cardiogenic shock with ductal closure
 - Left atrial hypertension and resultant pulmonary edema and pulmonary hypertension with right ventricular dilatation
 - Preoperative management of shock in critical AS
 - Prostaglandin infusion
 - Vasoactive support as needed to increase contractility
 - Mechanical ventilation to reduce metabolic demand
 - Correct metabolic acidosis
 - Monitor for signs of end-organ compromise
 - In older children, chronic pressure load may lead to ventricular dilatation and decreased function
 - Subendocardial ischemia may result from progressive hypertrophy and abnormal coronary perfusion

Operative or Interventional Procedures

- Intervention indicated if gradient > 50 mmHg or presence of symptoms
- Surgical valvotomy, valvuloplasty or aortic valve replacement (size limitations in neonates)
- Balloon valvuloplasty of aortic valve
- Ross procedure considered for patients with severe aortic insufficiency
- Subaortic stenosis: resection of muscle or membrane, modified Konno procedure
- Supravalvar aortic stenosis: patch plasty, arch reconstruction or arch replacement

Postoperative Considerations and Management

- Left ventricle function usually preserved and often hyperdynamic due to long standing stenosis and high afterload which may result in hypertension
- Intraoperative myocardial protection may be challenging in hypertrophied heart, anticipate more severe and more prolonged low cardiac output syndrome
- Valvotomy/valvuloplasty may result in aortic insufficiency

- Positive pressure ventilation may be useful to therapy to reduce LV afterload and improve cardiac output in the setting of LV dysfunction
- Monitor arterial and central venous pressure, may require left atrial pressure monitoring in select cases (lesions associated with VSD or mitral valve disease)

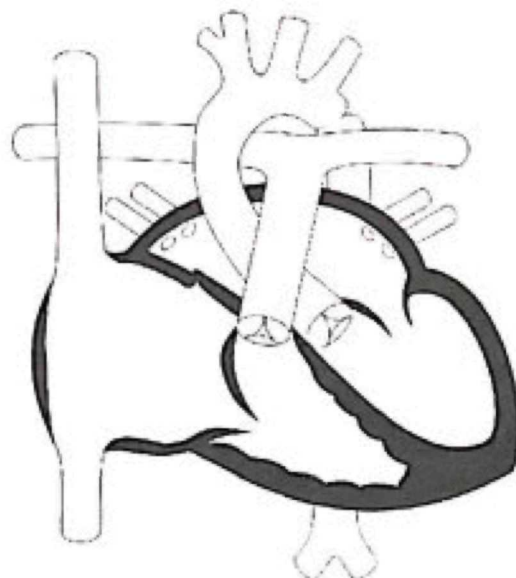
Lesion Specific Postoperative Concerns

- Anticoagulation if artificial valve
- Residual AS and/or aortic insufficiency
- Left bundle branch block (LBBB) and complete heart block in subaortic resection
- Coronary insufficiency

Section 2
Coarctation of the Aorta
Jeremy Affolter, MD

Anatomy

- Thoracic aorta narrowing, commonly juxtaductal with varying degrees of arch hypoplasia
- Associated cardiac defects
 - Bicuspid aortic valve +/- stenosis
 - Subaortic stenosis
 - Mitral valve stenosis
 - Ventricular septal defect
 - Hypoplasia of aortic valve, mitral valve, and left ventricle
- Common syndromes
 - Shone's complex
 - Hypoplastic left heart syndrome
 - Turner syndrome



Coarctation of the Aorta

Preoperative Physiology

- Neonatal presentation differs from disease presenting in childhood
- Critical coarctation associated with ductal dependent systemic blood flow
- Closure of the ductus arteriosus may result in inadequate systemic blood flow
 - May result in metabolic acidosis, organ dysfunction, and circulatory failure
- Acute rise in LV afterload
 - Poor compliance results in low cardiac output and pulmonary edema

- Presence of a VSD is protective against acute increase in LV afterload
- Childhood disease presents with left ventricular hypertrophy, thoracic aortic collaterals, and systemic hypertension
- Management
 - Establish prostaglandin infusion
 - Establish reliable venous access and arterial pressure monitoring.
 - Consider mechanical ventilation to decrease metabolic demand and LV afterload
 - Consider vasoactive support for ventricular dysfunction and organ support
 - End-organ assessment (neurologic status, renal and hepatic function, coagulopathy, GI tract)
 - Sepsis evaluation

Operative or Interventional Procedures

- Timing dependent on severity of coarctation and age at presentation
- When CPB required, may elect to wait for end-organ recovery
- Discrete coarctation approached through left thoracotomy without CPB
 - Complex coarctation/arch hypoplasia may require median sternotomy +/- CPB
- Coarctation resection and end-to-end anastomosis
 - Includes ductal tissue resection
 - Modifications including end-to-side and interdigitated repairs may reduce re-coarctation rates
- Subclavian flap angioplasty
 - Low re-coarctation rate
 - Diminished right arm blood flow
 - Concern for long term subclavian steal syndrome
- Patch aortoplasty
 - Higher re-coarctation rate and aneurysm formation
- Coexistent VSD includes
 - Primary VSD repair
 - Pulmonary artery banding
 - No surgery with medical management of congestive heart failure
- Balloon angioplasty +/- stent placement
 - Uncommon as an initial procedure in infancy
 - More commonly used in recurrent coarctation after surgery

Postoperative Considerations and Management

- Decreased afterload on LV, increase in cardiac output, and improved ventricular compliance
- Degree of preoperative ventricular and other organ dysfunction may impact postoperative course

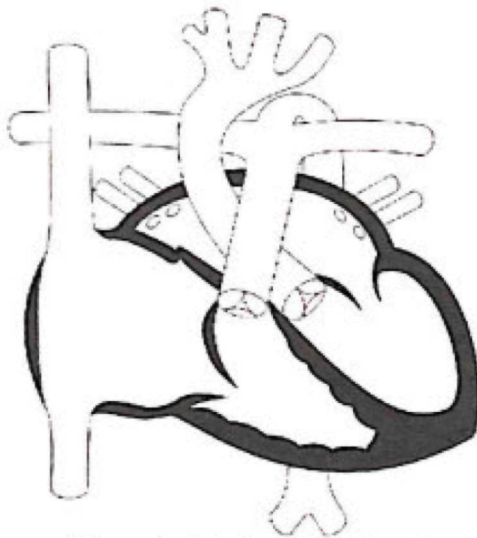
Lesion Specific Postoperative Concerns

- **Systemic hypertension**
 - Less common in infants
 - Risk for bleeding at aortic suture lines
 - Hypertension treatment associated with reduced incidence and severity of post-coarctectomy syndrome
- **Post-coarctectomy syndrome**
 - Most common in older patients with collateralization of aorta
 - Reperfusion injury to gastrointestinal tract causing bowel ischemia
 - May be related to uncontrolled hypertension
 - Conservative treatment until symptoms subside (gastric decompression, bowel rest, treatment of hypertension)
- **Residual coarctation**
 - Residual gradient > 20 mmHg
 - Gradient may not be evident initially if low cardiac output is present
 - Need for second intervention based on clinical course and timing after surgery
- **Residual lesions (i.e. VSD)**
 - May require medical or surgical intervention if associated with significant heart failure symptoms
- **Injury to structures in proximity to repair**
 - Thoracic duct leading to chylothorax
 - Recurrent laryngeal nerve leading to vocal cord dysfunction
 - Phrenic nerve injury leading to diaphragm dysfunction
- **Spinal cord injury from spinal artery ischemia**
 - Confirm baseline neurologic examination

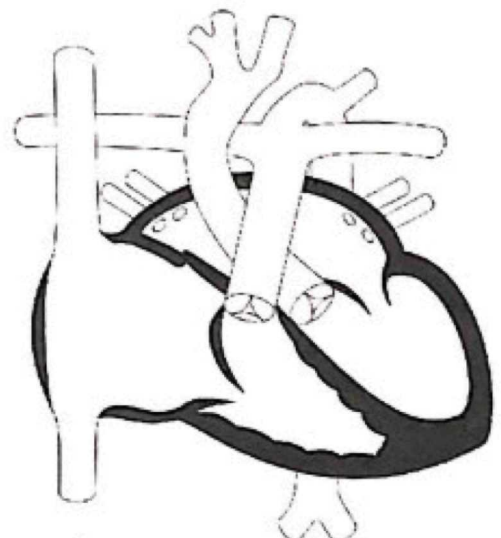
Section 3
Interrupted Aortic Arch
Shanelle Clark, MD

Anatomy

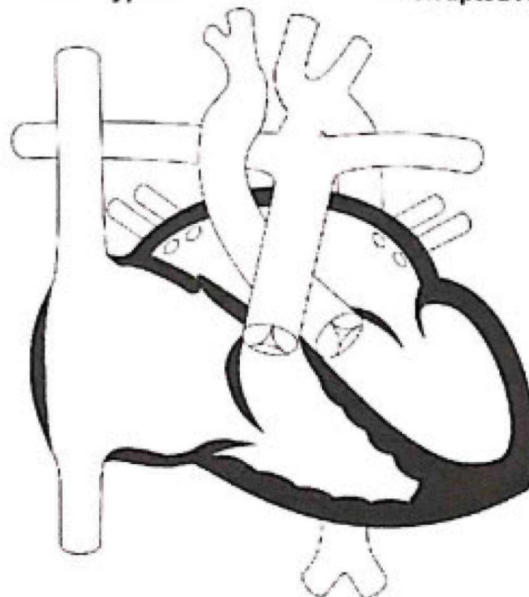
- Complete interruption of ascending and descending aorta
- 3 distinct type
 - Type A: interruption distal to the left subclavian artery at the level of the aortic isthmus
 - Type B (most common): interruption between the left carotid and left subclavian artery
 - Type C: interruption between the right innominate and the left carotid artery



Interrupted Aortic Arch - Type A



Interrupted Aortic Arch - Type B



Interrupted Aortic Arch - Type C

- Associated lesions
 - VSD
 - Left ventricular outflow tract obstruction (LVOTO)
 - Due to a posterior malalignment of the conal septum
 - Aortic valve hypoplasia, stenosis
 - Bicuspid aortic valve
 - Aberrant right subclavian artery (usually Type B)
 - Can occur with truncus arteriosus, transposition of the great arteries, and aortopulmonary window
- Association with 22q11.2 deletion syndrome
 - Most commonly found in Type B
 - May impact postoperative management (hypocalcemia, immune deficiency, neurologic, and oropharyngeal abnormalities)

Preoperative Physiology

- Circulatory shock and congestive heart failure may occur
 - Ductal dependent systemic blood flow necessitates initiation of prostaglandin
 - Shock presentation in late diagnosis or if ductus closes
 - May require resuscitation, mechanical ventilation, and inotropic support
 - End-organ damage such as acute kidney injury or necrotizing enterocolitis may result from inadequate perfusion
 - Combination of LVOTO and large VSD may lead to early pulmonary over-circulation and congestive heart failure
 - Avoid excessive FiO_2 and hyperventilation

Operative Procedure

- Median sternotomy with cardiopulmonary bypass
 - Aortic arch reconstruction
 - Direct anastomosis may require extensive patch augmentation of the arch in an end-to-side fashion
 - Interposition graft is rarely used
 - VSD closure by trans-atrial or trans-ventricular technique
 - Relief of aortic stenosis and/or subaortic obstruction
 - May require a Ross-Kono procedure, or a Norwood-type arch reconstruction combined with RV-PA conduit placement

Postoperative Considerations and Management

- Severity of low cardiac output syndrome dependent on total support time and cardiopulmonary bypass strategy
- Potential for elevated pulmonary vascular resistance in the setting of increased pulmonary blood flow preoperatively
- Prolonged left ventricular support particularly in patients with preoperative shock or left ventricular dysfunction

- Arrhythmia and/or heart block most likely when associated with VSD repair

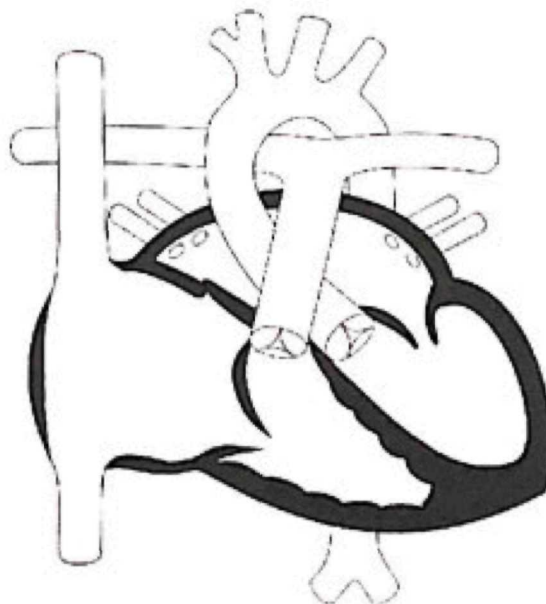
Lesion Specific Postoperative Concerns

- Residual or recurrent aortic coarctation
- Left ventricular outflow tract obstruction
 - Risks for development include a smaller aortic root size and aortic annulus size
- Residual or additional VSD
 - Significant shunt: heart failure symptoms necessitating continued heart failure management or possible re-intervention
- Left pulmonary artery and/or bronchial compression
 - Very rare
 - More common following direct anastomosis with inadequate mobilization of the aorta
- High risk of recurrent laryngeal nerve injury
 - Vocal cord paralysis may impact ability to extubate
- Risk of phrenic nerve injury
 - Hemidiaphragm paresis may impact ability to extubate
 - More commonly seen with a descending aortic conduit
- Hypocalcemia typically associated with 22q11 deletion

Section 4
Mitral Valve Stenosis
Richard Speicher, MD and
Aimee Jennings RN, CPNP

Anatomy

- Mitral apparatus consists of the anterior and posterior leaflets, chordae tendinae, and two discrete papillary muscles
- Congenital mitral valve disease is rarely an isolated abnormality
- May be associated with more complex disease
 - Hypoplastic left heart syndrome
 - Shone's complex
- Supravalvar mitral stenosis
 - Supravalvar mitral ring
 - Cor triatriatum: discrete left atrial membrane which obstructs mitral inflow
- Valvar mitral stenosis
 - Hypoplastic annulus
 - Leaflet fusion
 - Double orifice
- Subvalvar mitral stenosis
 - Parachute: all chordae funneling into single papillary muscle
 - Arcade: fused and shortened chordae



Mitral Stenosis

Preoperative Physiology

- Elevated left atrial pressure and volume
 - Elevated pulmonary venous and pulmonary arterial pressure
 - Pulmonary hypertension may develop

- Symptoms depend on degree and duration of mitral stenosis
 - tachypnea, wheezing, and failure to thrive
 - low cardiac output in severe cases
- Atrial arrhythmias may occur particularly if stenosis is long standing
- Preoperative management
 - Diuretics for pulmonary venous congestion
 - Digoxin and beta blockade may offer marginal benefit
 - Arrhythmia management
 - Severe disease may result in respiratory failure requiring intubation

Operative or Interventional Procedures

- No consensus indications for intervention exist: may include pulmonary hypertension, refractory right heart failure, inability to wean from positive pressure ventilation or rapid progression of gradient
- When possible, preservation of native mitral apparatus preferable to replacement
- Repair may require addressing multiple levels of obstruction and may include
 - Commissurotomy
 - Leaflet resection
 - Papillary muscle splitting or relocation
 - Chordae fenestration, shortening, or transfer
 - Supravalvar mitral ring resection
- Mitral valve replacement
 - Mechanical valve
 - Limited by size and need for anticoagulation
 - Higher mortality than repair
 - Risks include thrombosis, stroke, heart block, endocarditis
 - Implantation in annular or supra-annular position determined by annular size and left atrial size
- Balloon dilation
 - Limited to specific mitral valve morphologies
 - May result in increased insufficiency

Postoperative Considerations and Management

- Physiology dictated by presence and severity of residual stenosis and/or insufficiency
- Anticipate pulmonary hypertension
- Maintenance of adequate left atrial pressure and left ventricular preload
- Management of atrial arrhythmia
- Low cardiac output may be significant if mitral stenosis is associated with preoperative mitral valve regurgitation which provides volume load and decompression of left ventricle preoperatively

- Prolonged mechanical ventilation may be in presence of residual mitral stenosis or regurgitation and left ventricular dysfunction

Lesion Specific Postoperative Concerns

- Atrial arrhythmias and/or heart block
- Avoid volume overload which can cause left atrial distension
- Minimize hypertension and tachycardia to allow adequate diastolic filling time
- Pulmonary hypertension
 - Long standing preoperative stenosis
 - Supraaortic position of prosthetic valve may limit left atrial size and decrease filling
 - May cause significant right ventricular dilatation resulting in left ventricular compression
- Left atrial hypertension
 - Residual mitral stenosis
 - Residual mitral regurgitation
 - Left ventricular dysfunction
- Anticoagulation for bioprosthetic and mechanical prosthetic valves

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CHAPTER 7

Cyanotic Lesions

Chapter Editors

Josh Wolovitz, MD

Andrea Torzone, RN, MSN, CPNP, CNS

Section 1: Pulmonary Stenosis

Section 2: Tetralogy of Fallot

Section 3: Tetralogy of Fallot: Pulmonary Atresia

Section 4: Tetralogy of Fallot: Absent Pulmonary Valve

Section 5: Pulmonary Atresia: Intact Ventricular Septum

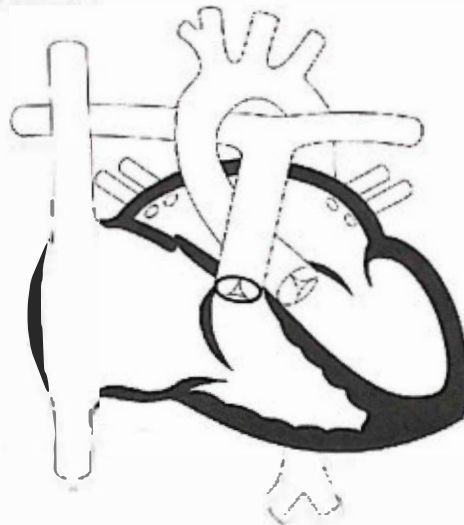
Section 6: Ebstein's Anomaly

Section 7: Total Anomalous Pulmonary Venous Return

Section 1
Pulmonary Stenosis
Dipankar Gupta, MD, F. Jay Fricker, MD and
Jai P. Udassi, MD, FACC, FAHA

Anatomy

- Valvar, subvalvar, and/or supra-valvar stenosis; MPA and/or branch stenosis
 - Classic valvar stenosis is conical or dome shaped with raphe and leaflet fusion
 - Dysplastic valvar stenosis with immobile and thickened leaflets
Noonan's syndrome
- Associated cardiac defects
 - Branch pulmonary artery stenosis
 - Ventricular septal defect
 - Tetralogy of Fallot
- Common syndromes
 - Noonan's syndrome
 - William's syndrome
 - Alagille syndrome



Pulmonary Stenosis

Preoperative Pt

- **Presentation**
 - Critical pulmonary stenosis in neonates associated with ductal dependent pulmonary blood flow
 - Newborns with severe stenosis present with suprasystemic RV pressure, right to left atrial shunting, hypoxemia, and acidosis as ductus arteriosus closes
 - Associated infundibular hypertrophy results in dynamic subvalvar obstruction, post-stenotic dilation of MPA and branch PA's
 - Progressive stenosis leads to RV dilation, decreased function, and tricuspid regurgitation
 - Mild cases may go undetected and present as murmurs, exertional dyspnea

- Management
 - Establish PGE infusion with critical neonatal stenosis
 - Assessment of valve leaflets, annular size and gradient at pulmonary valve by 2-D echocardiogram guides intervention

Operative or Interventional Procedures

- Candidates: RV pressure > 50% systemic, symptomatic patient, right ventricular dysfunction
- Balloon pulmonary valvuloplasty
- Surgical valvuloplasty through median sternotomy
- Patch plasty for subvalvar and supralvalvar stenosis
- Associated VSD closure with Dacron or autologous pericardium

Postoperative Considerations and Management

- Right ventricular dysfunction and decreased compliance
- Close attention to volume status, often require fluid resuscitation, avoid hypovolemia
- May need inotropic support immediately after weaning from CPB
- Inotropic support can be weaned off after 12-24 hours postoperatively
- Usually able to wake from anesthesia and wean from mechanical ventilator support with 24 hours
- Prolonged mechanical support may be needed in the setting of persistent complications (arrhythmias or bleeding)
- Maintain adequate preload to support noncompliant right ventricle

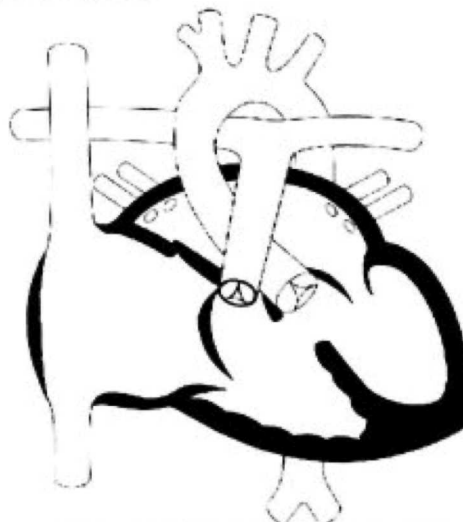
Lesion Specific Postoperative Concerns

- Right ventricular noncompliance
 - Persistent cyanosis due to RV noncompliance and right to left atrial shunt
 - Predisposes to arrhythmia
 - Suicide right ventricle (severe RV contractility failure due to severe right ventricular outflow tract obstruction)
 - Support with inotropy and adequate preload
- Residual valvar stenosis/regurgitation
 - Audible systolic murmur radiating to back and axilla with stenosis
 - Diastolic high pitched murmur at left upper sternal border with pulmonary regurgitation
 - Significant stenosis may precipitate or prolong RV dysfunction
- Arrhythmia
 - Junctional ectopic tachycardia, atrial tachycardia, ventricular tachycardia
 - Associated with VSD closure and/or RV muscle resection

Section 2
Tetralogy of Fallot
Roxanne Kirsch, MD, FRCPC, FAAP

Anatomy

- Overriding aorta (over RV) with anterior malaligned VSD, subpulmonary and/or pulmonary valve obstruction, and RV hypertrophy
- Associated cardiac defects
 - Coronary artery abnormalities (5% of coronary anomalies have LAD from RCA crossing subpulmonary outflow tract)
 - TOF with complete AV canal defect
 - Anomalous pulmonary venous drainage
 - Right aortic arch (25%)
 - Left superior vena cava
- Common syndromes
 - 22q11 deletion
 - Trisomy 21
 - Alagille Syndrome
 - Pentalogy of Cantrell



Tetralogy of Fallot with Pulmonary Stenosis

Preoperative Physiology

- Cyanosis
 - Dependent on amount RV outflow tract obstruction and pulmonary valve stenosis
 - Hypercyanotic spells caused by obliteration of pulmonary blood flow
 - Lack ejection systolic murmur
- Congestive heart failure
 - Qp/Qs dependent on amount of pulmonary stenosis
 - VSD usually unrestrictive

Operative Procedure

- Full repair in neonatal period or electively at 3-6 months
- Patch closure of VSD and relief of RV outflow tract and pulmonary valve obstruction

- VSD closure via right atriotomy through the tricuspid valve, may be approached through infundibulotomy or ventriculotomy
- RV muscle bundle resection, transannular patch (which might extend to one or both branch PA), infundibular patch, infundibular and MPA patch, pulmonary valvotomy
- RV-PA conduit may be required due to LAD from RCA crossing RVOT
- May be palliated with BT shunt as a neonate with full repair in early infancy

Postoperative Consideration and Management

- May need inotropic support immediately after weaning from CPB to support right ventricle
- Lusitropic agents such as milrinone aid with diastolic dysfunction
- Typically inotropic support can be weaned off after 12-24 hours postoperatively
- Usually able to wake from anesthesia and wean from mechanical ventilator support with 24 hours
- Mechanical ventilation with high mean airway pressure will exacerbate RV dysfunction
- When hemodynamically possible, spontaneous breathing will decrease RV afterload and improve right ventricular filling
- Prolonged mechanical support may be needed in the setting of persistent complications (arrhythmias or bleeding)
- Degree of right ventricular diastolic dysfunction will dictate fluid management
- Must ensure adequate right ventricular preload to maintain cardiac output
- Diuretics will be needed postoperatively, may be delayed if ongoing fluid resuscitation necessary for RV dysfunction

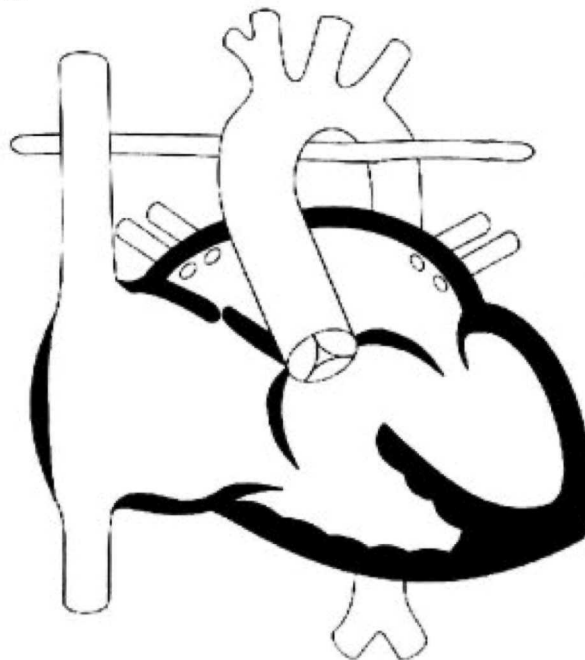
Lesion Specific Postoperative Concerns

- Right ventricular noncompliance
 - Most common in patients with significant outflow tract obstruction and right ventricular hypertrophy
 - Residual RV outflow obstructions will worsen this phenomenon
 - Presents with low cardiac output, persistent tachycardia and high right atrial pressure
 - Cyanosis usually result of persistent right to left atrial shunt
 - Absence of atrial septal defect will lead to profound low cardiac output with normal saturation
- Arrhythmia
 - Junctional ectopic tachycardia is commonly associated with TOF repair
 - May result from noncompliant RV, hematoma, edema, and suture line along VSD patch
 - Generally resolved within 24-72 hours postoperatively and does not recur
 - Complete heart block
- Residual VSD
 - Usually hemodynamically significant
 - Left atrial pressure greater than right atrial pressure
 - Associated with persistent tachycardia, inability to achieve adequate diuresis, prolonged duration of mechanical ventilation, and congestion on chest x-ray

Section 3
Tetralogy of Fallot Pulmonary Atresia
Kasey Zawodniak, MSN, RN, PCCNP

Anatomy

- Atresia of the pulmonary valve with anterior malaligned VSD (may also be membranous or infundibular)
- Wide range of origin, size, and distribution of pulmonary blood flow
 - PAs well developed, confluent and blood flow supplied by large PDA
 - PDA small, PAs small (< 2 mm) with a few major aorta to pulmonary collaterals (MAPCAs)
 - Discontinuous PAs with variable blood supply
 - No true PAs or PDA, all blood supplied through MAPCAs
- Absence or severe hypoplasia of the true PAs most severe type of disease and increases morbidity
- Moderate to severe right ventricular hypertrophy (RVH)
- Right aortic arch 25-50%
- ASD/PFO 50%



Tetralogy of Fallot - Pulmonary Atresia

Preoperative Physiology

- Obligatory right to left shunt through VSD (and ASD if present)
- Pulmonary blood flow is dependent on PDA and/or MAPCAs
- Clinical presentation is dependent upon pulmonary blood flow
 - Cyanosis if ductal-dependent

- MAPCAs may provide adequate pulmonary blood flow (PBF) until infant outgrows relatively fixed PBF
- Large PDA or multiple MAPCAs may contribute to symptoms of CHF due to significant left to right shunting
- Cardiac catheterization
 - Determine PA anatomy including additional sources of PBF via MAPCAs or PDA
 - Delineate size and distribution of true PAs and MAPCAs
 - Evaluate for multiple VSDs
 - Aortic arch anatomy

Operative or Interventional Procedures

- Operative procedure is dependent upon PA anatomy
- Ductal-dependent PBF and confluent PAs
 - Primary repair with RV-PA conduit and VSD closure
 - Staged with BTS or PDA stent, followed by similar repair when PA growth is adequate and there is evidence of good arborization of the pulmonary vascular bed
- MAPCAs
 - Goal is to promote growth of the hypoplastic PAs by increasing PBF
 - Staged palliation with ultimate goal of complete repair
 - Unifocalization of MAPCAs and true PAs
 - Modified BT shunt to hypoplastic true pulmonary arteries or conduit from RV to unifocalized MAPCAs and/or true hypoplastic PAs
 - Coil embolization of superfluous collaterals
 - Surgical ligation of collaterals
 - Coil embolization in catheterization lab

Postoperative Consideration and Management

- Usually inotropic support can be weaned off after 12-24 hours postoperatively
- Usually able to wake from anesthesia and wean from mechanical ventilator support within 24-48 hours
- Prolonged mechanical support may be needed in the setting of persistent complications: arrhythmia, bleeding, MAPCAs or hypoxemia

Lesion Specific Postoperative Concerns

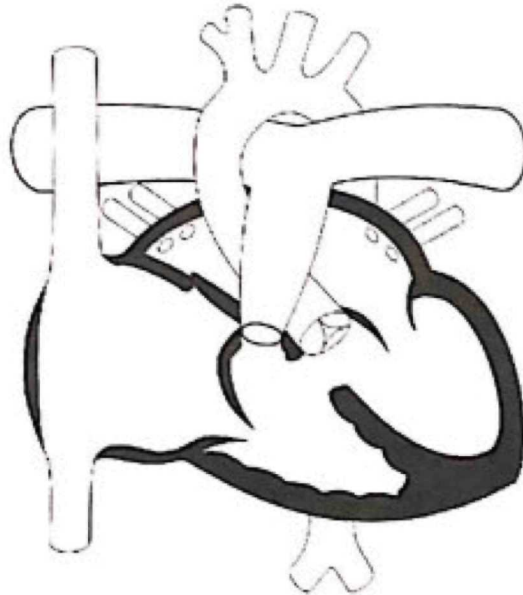
- Arrhythmias
 - Secondary to edema or suturing
 - JET and complete heart block most common
- RV dysfunction
 - May require volume resuscitation and higher CVP due to noncompliant RV

- Avoid tachycardia to improve preload of RV
- Decrease right ventricle afterload to promote pulmonary blood flow
- Rapid extubation if possible to lower intrathoracic pressure and decrease RV afterload
- Consider addition of i O in cases of persistent hypoxemia
- Residual VSD
 - When trivial or small, typically not hemodynamically significant
 - May cause persistent left to right shunting, LA dilation, and heart failure symptoms due LV volume load
 - Often associated with persistent tachycardia, inability to achieve adequate diuresis, and prolonged duration of mechanical ventilation

Section 4
Tetralogy of Fallot Absent Pulmonary Valve
Sherrie Joy Baysa, MD

Anatomy

- Undeveloped pulmonary valve leaflets: stenotic and regurgitant
- Aneurysmal, massive dilatation of pulmonary arteries secondary to in-utero pulmonary regurgitation
- Intracardiac features of TOF
- Should be considered a syndrome with associated findings in the lungs and airways
 - Associated tracheobronchial anomalies including significant malacia
 - Abnormal pulmonary vascular branching and wall structure



Tetralogy of Fallot - Absent Pulmonary Valve

Preoperative Physiology

- Severe pulmonary valve regurgitation in-utero leads to hydrops in up to 20%
- Present as neonates with cyanosis and a harsh to-and-fro murmur of pulmonary stenosis and regurgitation
- Many have associated respiratory distress/failure secondary to bronchial compression from the PAs and abnormal distal arteriolar/alveolar architecture leading to V/Q mismatch
- In those with respiratory distress and cyanosis
 - Prone positioning to decrease airway compression occasionally helpful
 - With frank respiratory failure, intubate and initiate mechanical ventilation
 - Higher than normal PEEP to stent airways open
- High prevalence of 22q11 deletion
- Patients with mild disease present with TOF physiology

Operative Procedure

- Usually require intervention in the newborn period
- Repair through an infundibulotomy and transannular incision
 - VSD closure and relief of RVOT obstruction
 - Anterior plication of the PAs to reduce airway compression
 - Possible excision of dilated PAs with placement of a homograft
- Mortality increases in neonates with respiratory failure

Postoperative Consideration and Management

- Often require prolonged mechanical ventilation for persistent respiratory insufficiency secondary to malacia and ventilation perfusion mismatch
- Delayed sternal closure may help alleviate airway compression immediately postoperatively
- May liberalize fluid administration to augment preload in the setting of diastolic dysfunction
- Respiratory and RV hemodynamics dictate depth and duration of sedation

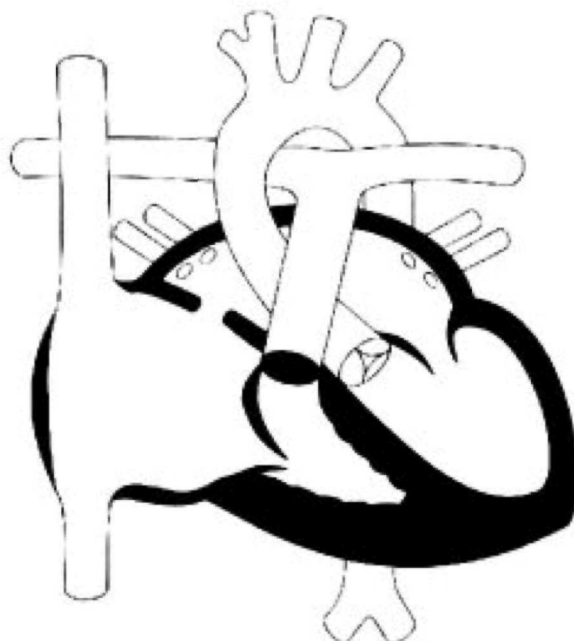
Lesion Specific Postoperative Concerns

- Continued bronchial obstruction
 - Residual airway hypoplasia and abnormal distal architecture
 - Improves with time as PA pressures decrease and the tracheobronchial tree matures/become less compressible
 - Commonly have long standing respiratory insufficiency and may require tracheostomy placement for chronic ventilation
- RV dysfunction
- Pulmonary regurgitation and progressive RV dilation
 - Close surveillance of degree of RV dilation and/or RV dysfunction
 - May necessitate pulmonary valve replacement

Section 5
Pulmonary Atresia with Intact Ventricular Septum
Julie Wells, MSN, RN, CPNP and
Maria Teresa Zuluaga, MD

Anatomy

- Imperforate pulmonary valve
- Ventricular septum functionally intact
- Hypertrophic RV with normal to hypoplastic cavity
- Tricuspid valve diameter correlates with RV cavity size
- Dilated RA with PFO or ASD
- May have ventriculo-coronary sinusoids, coronary stenosis, or right ventricular-dependent coronary circulation



Pulmonary Atresia with Intact Ventricular Septum

Preoperative Physiology

- Complete obstruction to RV outflow leads to moderate to severe tricuspid regurgitation and marked right atrial dilation
- Right to the left atrial shunt
- Pulmonary blood flow dependent on patent ductus arteriosus
- In some patients the hypertensive RV drives blood through sinusoids within the myocardium to the coronary arteries
- Saturations should guide adequacy of pulmonary blood flow as in preoperative single ventricle patients
- Goals of management
 - Restore or maintain ductal patency with prostaglandin
 - Treat hypoxia, acidosis, and shock when present

- Despite obligate right to left atrial shunt, patients may require balloon atrial septostomy
- All patients must have preoperative cardiac catheterization to evaluate hemodynamics, RV structure and size, and coronary artery anatomy
 - Right ventricular dependent coronary circulation (RVDCC) is defined by the presence of proximal coronary artery stenosis and coronary blood flow is dependent upon flow from hypertensive right ventricle

Operative and Interventional Procedures

- **Biventricular repair**
 - Usually associated with mild to moderate RV hypoplasia
 - RVDCC must be excluded before opening the pulmonary valve and decompressing the RV
 - Must relieve obstruction to pulmonary blood flow
 - Catheter based balloon valvuloplasty
 - Surgical RVOT augmentation
 - Modified BT shunt or prostaglandins to maintain patency of PDA indicated in patients with persistent cyanosis following relief of pulmonary artery stenosis
 - PFO/ASD is left open
- **Single ventricle pathway**
 - Severe RV hypoplasia and/or RVDCC
 - Modified BT shunt or PDA stent placement to provide pulmonary blood flow
 - Patients with RVDCC and signs of coronary ischemia may require early evaluation for heart transplantation

Postoperative Considerations and Management

- Dependent on interventional/surgical pathway
- Must have high index of suspicion for coronary insufficiency if any arrhythmias or ECG changes are noted
 - Reevaluate for the presence of RVDCC

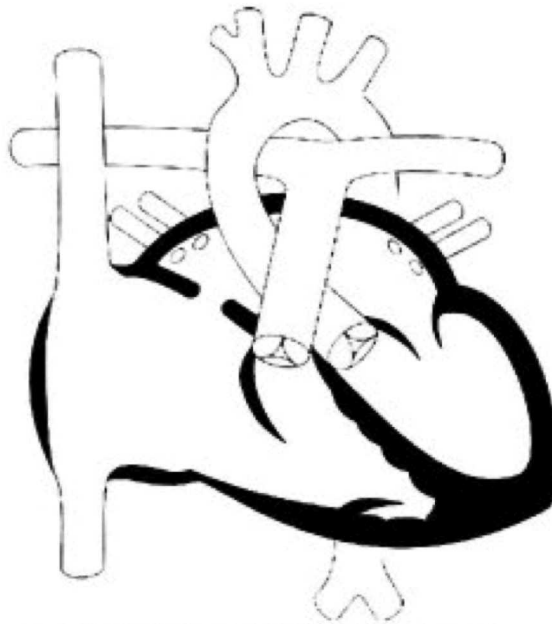
Lesion Specific Postoperative Concerns

- Circular shunt physiology
 - In patients with a transannular patch for relief of RVOT obstruction and a PDA or modified BT shunt
 - During diastole, blood flows from shunt or PDA retrograde into the RV across the regurgitant pulmonary valve
 - During systole, blood flows retrograde into the RA in the presence of tricuspid regurgitation
 - Oxygenated blood from the LV and aorta return to the RA and results in systemic hypoperfusion and inadequate oxygen delivery

- Profound low cardiac output result: hypotension, oliguria, and metabolic acidosis
- Management
 - Increase pulmonary vascular resistance and decrease systemic vascular resistance
 - Usually requires surgical intervention to narrow shunt and/or decrease tricuspid regurgitation
- Cyanosis
 - Usually results from combination of RV dysfunction, persistent RVOT obstruction, and right to left atrial shunt
 - Saturations should be used to gauge adequacy of pulmonary blood flow
 - Decrease right ventricle afterload to promote pulmonary blood flow
 - Rapid extubation if possible
 - Minimize mean airway pressure
 - Consider addition of iNO in cases of persistent hypoxemia
 - RV dysfunction due to right ventriculotomy
 - Right to left shunt allows preservation of cardiac output
 - Patients may be cyanotic while awaiting recovery of RV diastolic function
- Arrhythmia
 - Evaluate for RVDCC

Anatomy

- The posterior and septal leaflets of the tricuspid valve are displaced downward into the right ventricle and is associated with tricuspid regurgitation
- Right ventricular outflow tract obstruction can be caused by pulmonary valve stenosis or obstruction from the "sail-like" anterior leaflet of the tricuspid valve
- The inlet portion of the RV is incorporated into the right atrium, causing atrialization of the RV and functional hypoplasia
- Commonly associated with ASD, PDA, and varying degrees pulmonary stenosis and/or atresia
- Associated with Wolff-Parkinson-White (WPW) type accessory pathways



Ebstein's Anomaly of the Tricuspid Valve

Preoperative Physiology

- The severity of symptoms can range from severe cyanosis and cardiovascular collapse in the newborn to nearly asymptomatic patients
- Cardiomegaly may cause airway compression and/or lung hypoplasia
- Congestive heart failure
 - Caused by combination of tricuspid valve regurgitation and severity of right ventricular hypoplasia and atrialization
 - Despite functional hypoplasia, right ventricle often markedly distended which may compromise filling of the left ventricle

- **Cyanosis**
 - Caused by combination of obstruction to pulmonary blood flow from pulmonary stenosis, obstruction from the anterior leaflet of the tricuspid valve with or without associated lung hypoplasia
 - Severe pulmonary regurgitation in patients with severe tricuspid regurgitation and patent ductus arteriosus may cause a circular shunt and severe cardiogenic shock in presence of cyanosis
 - Exacerbated by large right to left atrial shunt
 - May improve with decrease in pulmonary vascular resistance
 - Patients should be supported with therapies to encourage pulmonary blood flow and decrease pulmonary vascular resistance
- Arrhythmias (supraventricular tachycardia, atrial flutter, atrial fibrillation) as a result of severe right atrial dilatation

Operative Procedures

- Operative strategy determined by right ventricular size and ability to support full cardiac output
- **Single ventricle palliation**
 - Newborns with severe RV dysfunction and/or hypoplasia and ductal dependent pulmonary blood flow
 - Involves placement of a modified BT shunt, over-sewing of the tricuspid valve with a pericardial patch, RA plication and when necessary, an atrial septectomy
- **One and a half ventricle repair**
 - This strategy is used in patients with borderline RV volume and allows the possibility of a two ventricle conversion at a later time
 - Right ventricular outflow tract obstruction is relieved, the tricuspid valve is repaired with a right atrial plication to limit RV volume load
 - A bidirectional Glenn shunt is created to augment pulmonary blood flow
- **Two ventricle repair**
 - Performed in patients with adequate RV volume
 - The tricuspid valve is repaired or replaced with right atrial plication and ASD closure

Postoperative Consideration and Management

- **Low cardiac output**
 - May be exacerbated by both atrial and ventricular arrhythmias
 - Residual RV dysfunction or hypoplasia
 - Tricuspid insufficiency preoperatively causes RV dilation and an underfilled and poorly conditioned LV
- Pulmonary insufficiency as a result of airway compression or lung hypoplasia
- Inotropic support and afterload reduction for both ventricles will likely be needed for several days
- Both atrial and ventricular arrhythmias are common and may require electrical and pharmacologic management

- Full mechanical ventilation will likely be needed for several days
- Therapies should be focused on reducing pulmonary vascular resistance and maintaining lung expansion
- Weaning from positive pressure ventilation may decrease RV afterload and improve hemodynamics
- Prolonged diuretic therapy will be needed due to preoperative volume load and to optimize pulmonary V/Q matching
- Adequately controlling pain and agitation is essential to reduce oxygen demand and prevent an elevation in pulmonary vascular resistance

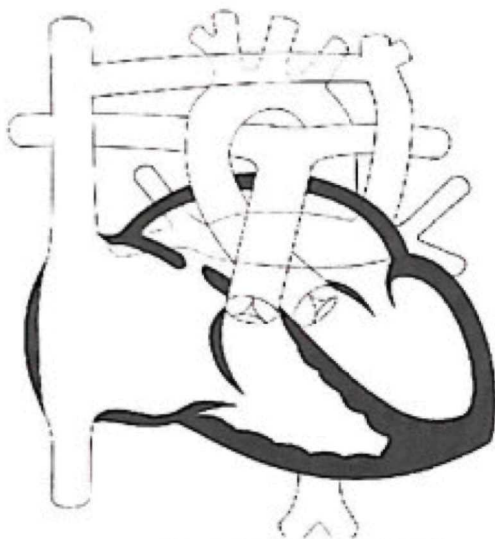
Lesion Specific Postoperative Concerns

- **Arrhythmias**
 - Often atrial tachycardias: atrial flutter, atrial fibrillation, and supraventricular tachycardia
 - Secondary to atrial dilation, residual tricuspid regurgitation, suturing or edema
 - Priority should be given to establishing atrioventricular synchrony
- **Residual tricuspid valve regurgitation**
 - May manifest as cyanosis and low cardiac output
 - Strategies to decrease pulmonary vascular resistance should be implemented
- **Ventricular dysfunction**
 - Right ventricular failure is common
 - Inotropic support and strategies to reduce right ventricular afterload must be considered
 - Left ventricular dysfunction is more likely in children with preoperative congestive heart failure
- **Respiratory failure**
 - Common in severe neonatal disease with severe cardiomegaly resulting in lung hypoplasia
 - Maintenance of adequate lung expansion with positive pressure ventilation and avoidance of overdistension

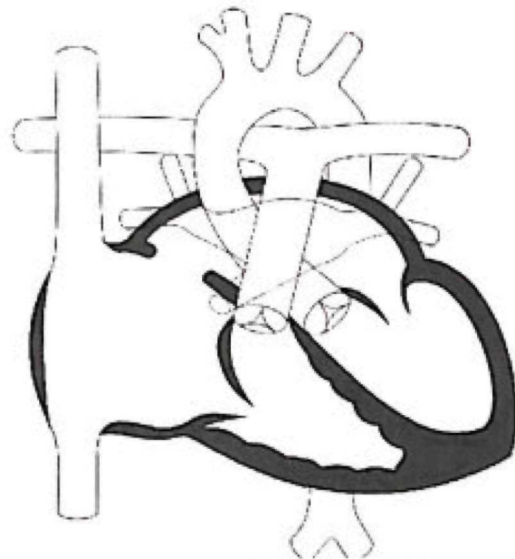
Section 7
Total Anomalous Pulmonary Venous Return
Jason Patregnani, MD

Anatomy

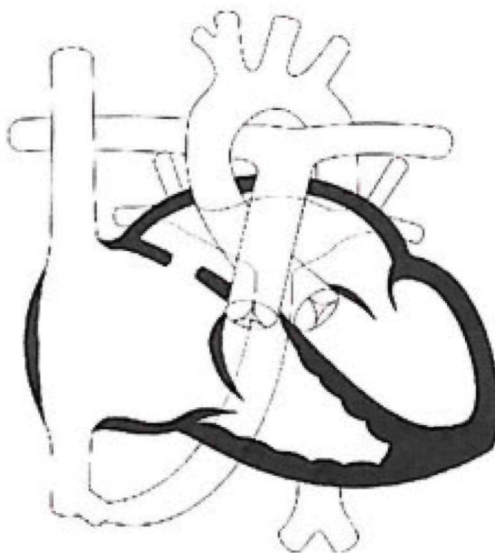
- Supracardiac (~45%): anomalous connection is to an ascending vertical vein, usually left sided and connected to the left innominate vein
- Cardiac (~25%): anomalous connection directly to the right atrium or coronary sinus
- Infracardiac (~25%): anomalous connection is to intra-abdominal veins, below the diaphragm
- Mixed (~5%): anomalous connection is mixed with at least one of the main lobar pulmonary veins connection to a different systemic vein
- Obstructed total anomalous pulmonary venous return (TAPVR)
 - Can occur with any subtype
 - Most commonly seen in infracardiac subtype



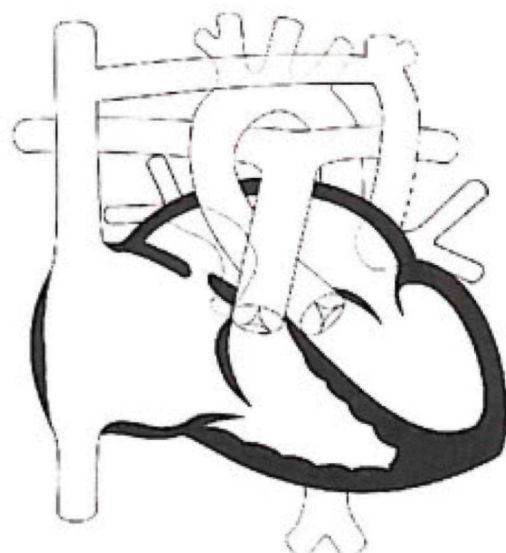
TAPVR - Supracardiac



TAPVR - Intracardiac



TAPVR - Infracardiac



TAPVR - Mixed type

Preoperative Physiology

- Unobstructed TAPVR
 - Both systemic and pulmonary venous return to the RA
 - Cardiac output is dependent on right to left atrial level shunt
 - Mixing of systemic and venous blood leads to variable levels of cyanosis determined by amount of pulmonary venous return, which depends on degree of pulmonary hypertension
 - Often leads to increased Qp/Qs as PVR drops over the first several weeks of life leading to heart failure symptoms
 - Often present with symptoms consistent with atrial septal defect
- Obstructed TAPVR
 - Chest x-ray with severe pulmonary venous congestion and small cardiac silhouette
 - Profound cyanosis with respiratory distress within minutes to hours after birth.
 - Often associated with metabolic acidosis
 - Preoperative management
 - Prostaglandin initiation can increase cardiac output (and increased mixed venous saturation) by allowing right to left shunting through ductus, but can decrease pulmonary blood flow and worsen cyanosis
 - Optimize pulmonary vascular resistance with intubation, correction of metabolic and respiratory acidosis as well as sedation and paralysis as needed
 - Consider ECMO prior to surgical intervention

Operative Procedure

- Anastomosis of pulmonary venous confluence to the left atrium on cardiopulmonary bypass

Postoperative Considerations and Management

- Patients with unobstructed type usually extubate within 24 hours and require minimal vasoactive support
- Pulmonary hypertension biggest concern for all types, but especially if obstructed preoperatively
- Low cardiac output syndrome secondary to
 - Small, noncompliant left ventricle and left-sided structures
- Obstructed TAPVR
 - Not uncommon to require ECMO
 - Often require prolonged inotropic support and lusitropic agents may assist with diastolic dysfunction
 - Supraventricular arrhythmias in up to 5-10% of patients
- Ventilator support necessary to manage respiratory insufficiency from preoperative pulmonary venous congestion, post-bypass inflammatory response
 - Positive end expiratory pressure helpful to optimize V/Q mismatch
 - iNO typically employed

- Often require prolonged sedation and/or neuromuscular blockade to optimize ventilation, allow for ventricular recovery, and minimize pulmonary hypertension

Lesion Specific Postoperative Concerns

- Low cardiac output
 - May result from relative LV noncompliance secondary to dilated right ventricle
 - May be exacerbated by postoperative pulmonary hypertension
- Pulmonary hypertension
 - Monitoring often assisted by presence of pulmonary artery line
 - Most common concern, especially following repair of obstructed TAPVR
 - Must rule out residual pulmonary venous obstruction
- Arrhythmia
 - 5-10% of patients have supraventricular arrhythmias
- Residual pulmonary venous obstruction
 - Persistence of typical CXR pattern of obstructed TAPVR
 - Persistent signs of low cardiac output

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CHAPTER 8

Transposition of the Great Arteries

Chapter Editors

David M. Axelrod, MD

Megan Tracey, RN, MS, PPCNP-BC, CNS

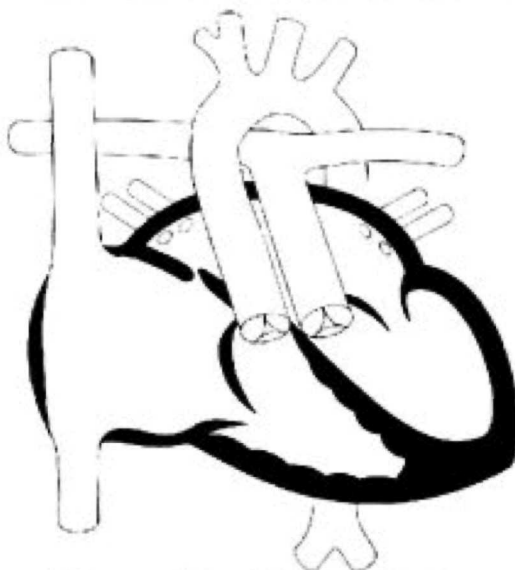
Section 1: D-transposition of the Great Arteries (D-TGA)

Section 2: L-transposition of the Great Arteries (L-TGA)

Section 1
D-transposition of the Great Arteries (D-TGA)
Julie Bushnell, RN, PNP, MS

Anatomy

- Situs solitus of atria, concordant atrial-ventricular connections with discordant ventricular-arterial connections
- Aorta arises from morphologic RV and pulmonary artery arises from morphologic LV
- Aorta rightward ("D" = "dextro" in D-TGA) and anterior to pulmonary artery
- Ventricular septal defect in approximately 25%; complex variants may have obstruction to pulmonary or systemic blood flow (i.e., Taussig-Bing anomaly)
- A number of coronary artery abnormalities may be present



D-Transposition of the Great Arteries

Preoperative Physiology

- Systemic and pulmonary circulations function in parallel, not in series
- Defining physiologic feature is pulmonary artery oxygen saturation greater than aortic oxygen saturation.
- Requires mixing at either the atrial level, ventricular level or at the level of the great vessels
 - Inadequate mixing leads to severe hypoxemia and acidosis
 - Decreased PBF or increased PVR can result in low arterial oxygen saturation despite adequate mixing
- Reverse differential cyanosis (i.e., hypoxemia with lower extremity O₂ sat > right upper extremity O₂ sat), is pathognomonic for D-TGA with pulmonary hypertension and/or aortic arch obstruction
- Pre and post ductal oxygen saturation monitoring may be used to detect reverse differential cyanosis
- Prostaglandin infusion may be required to promote flow from the aorta to the PA through the PDA to improve LA preload and atrial mixing
- A balloon atrial septostomy (Rashkind procedure) may be performed urgently to establish adequate atrial mixing if severely hypoxic or electively to allow for a trial of discontinuation of prostaglandin

Operative Procedure

- Arterial switch operation (Jatene procedure)
- Timing of surgical repair: often performed at 3-5 days of life to allow transition from fetal to neonatal circulation, end-organ recovery, and evaluation for additional congenital anomalies (uncommon in simple D-TGA)
- Technical details
 - CPB and aortic cross clamp
 - PDA divided and branch PAs exposed to provide mobility for anterior anastomosis
 - Great arteries transected above valvar level
 - Coronary arteries excised and reimplanted in neoaorta using "buttons"
 - LeCompte maneuver: PA is brought anterior to the aorta
 - Coronary donor sites filled with autologous pericardial patches
 - Pulmonary artery is reanastomosed just above neopulmonary valve

Postoperative Considerations and Management

- Afterload reduction and inotropic support may be needed to manage low cardiac output syndrome
- Avoid hypertension to minimize bleeding risk given aortic suture line
- Generally extubated within 48-72 hours

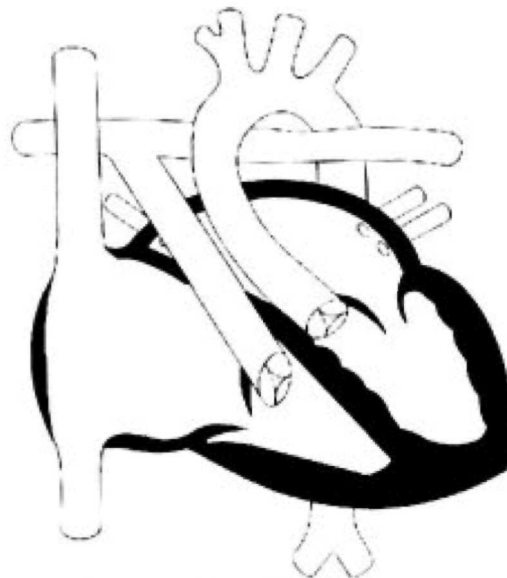
Lesion Specific Postoperative Concerns

- Coronary insufficiency resulting from coronary translocation and insertion
- LV dysfunction due to deconditioned LV (primarily seen in patients older than 6-8 weeks at time of surgery with intact ventricular septum and no PDA)
- Pulmonary hypertension

Section 2
L-transposition of the Great Arteries (L-TGA)
Stephanie L. Siehr, MD

Anatomy

- 95% situs solitus {S, L, L}
 - Ventricular inversion or L-looped ventricles
 - Atrioventricular and ventriculoarterial discordance
 - The aorta is positioned anterior/leftward of pulmonary artery
- 25% dextrocardia or mesocardia
- >90% have associated cardiac defects
 - VSD (80%)
 - Left ventricular outflow tract (LVOT) obstruction (30-50%)
 - Anomalies of the systemic tricuspid valve (TV) (20-50% Ebstein's-like)
- Abnormalities of atrioventricular node and bundle predispose to heart block
- Accessory connections (including WPW) may exist, allowing atrioventricular reciprocating tachycardia (SVT) to occur



L-Transposition of the Great Arteries

Preoperative Physiology

- Clinical features
 - Isolated L-TGA: may be asymptomatic for many years until systemic right ventricle (RV) begins to fail
 - Associated lesions determine symptoms and timing of presentation
 - Patients present earlier if inadequate pulmonary blood flow results in cyanosis, if VSD or TV anomalies result in heart failure, or if high grade atrioventricular block is present
- Management determined by associated lesions
 - Neonate with inadequate pulmonary blood flow requires prostaglandin infusion to maintain ductal patency
 - Systemic valve (left-sided tricuspid) regurgitation may lead to systemic ventricular dysfunction and congestive heart failure
 - VSD may result in congestive heart failure

Operative Procedures

- Operation depends on age, anatomy, and physiology
- Physiologic repair ("double-switch") to create atrioventricular (AV) and ventriculoarterial (VA) concordance
 - Atrial level switch (Mustard or Senning procedure)
 - Arterial switch (switching aorta and pulmonary artery with coronary transfer)
- Timing of double switch determined by associated lesions and projected readiness of left ventricle (LV) to tolerate systemic afterload following operation
- If the LV has become deconditioned from pumping against low afterload, a pulmonary artery (PA) band may be placed to condition the LV prior to a double switch
 - PA band may also improve tricuspid regurgitation by shifting the ventricular septum rightward
- Atrial switch plus Rastelli-type LVOT reconstruction with RV-PA conduit (instead of arterial switch) may be performed if there is severe PS or pulmonary atresia
- A modification of the double switch procedure is an arterial switch with a hemi-Mustard (IVC baffle to the left) with bidirectional Glenn to unload RV and reduce extent of atrial baffle
- Anatomic repair: leave AV/VA discordance and repair associated lesions
 - VSD closure
 - Tricuspid valve repair
 - Neonates with significant pulmonary stenosis (ductal dependent pulmonary blood flow) may initially need relief of obstruction or establishment of an alternative source of pulmonary blood flow (i.e., modified BT shunt)
- If no associated lesions may manage medically and monitor for systemic RV failure and atrioventricular valve regurgitation
- Pacemaker indicated if high grade 2nd degree or 3rd degree AV block
- Selected patients may ultimately be considered for cardiac transplantation

Postoperative Considerations and Management

- Post PA band
 - May need inotropic support for LV dysfunction
 - Postoperative extubation should be carefully planned due to tenuous LV function
- Post double-switch
 - Vasoactive support and afterload reduction may be needed to manage low cardiac output syndrome
 - May require temporary pacing for AV block
 - May require mechanical ventilation for the first 24-48hours

Lesion Specific Postoperative Concerns

- Post PA band: LV function usually depressed
- Post double-switch
 - LV dysfunction may result from acute increase in afterload
 - LV may be sensitive to volume load, diuretics indicated
 - Monitor for baffle leak or obstruction, residual LVOT obstruction

Risk of atrial arrhythmias given atrial baffle/suture lines/accessory connections

Risk of AV block warranting temporary or permanent pacing

CHAPTER 9

Single Ventricle Anatomy and Neonatal Palliation

Chapter Editors

Michael-Alice Moga, MD

Emily Petersen, RN, MS, CPNP

Section 1: Anatomy and Preoperative Physiology
of Common Single Ventricle Lesions

Section 2: Pulmonary Artery Band

Section 3: Blalock Taussig Shunt

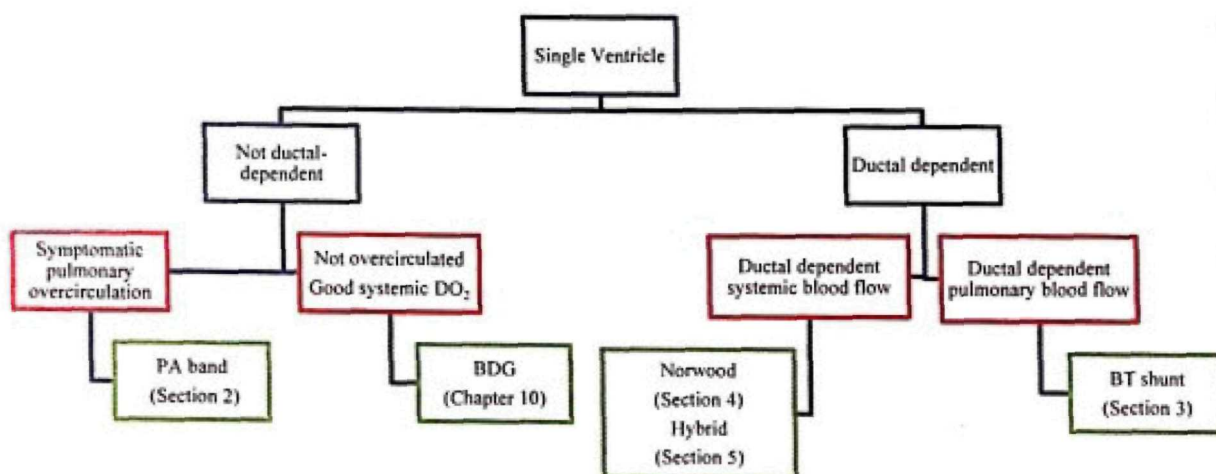
Section 4: Norwood Procedure

Section 5: Hybrid Procedure

Section 1

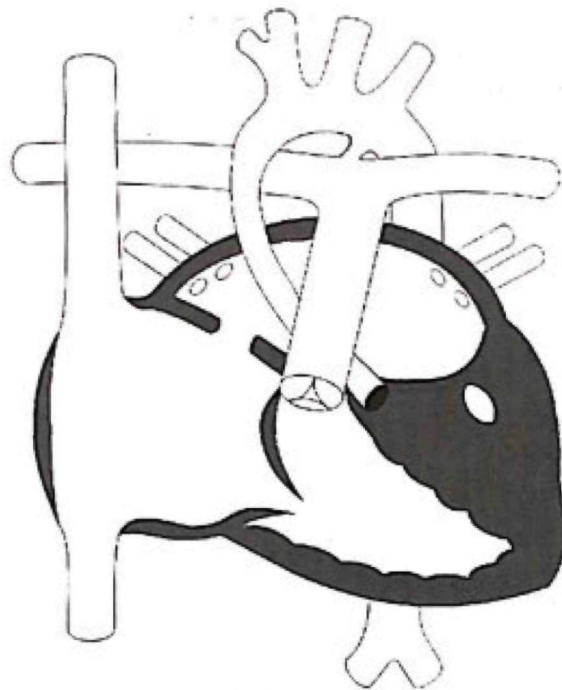
Anatomy and Preoperative Physiology of Common Single Ventricle Lesions

Robert Palermo, DO and Francesco Grasso, MD



Anatomy

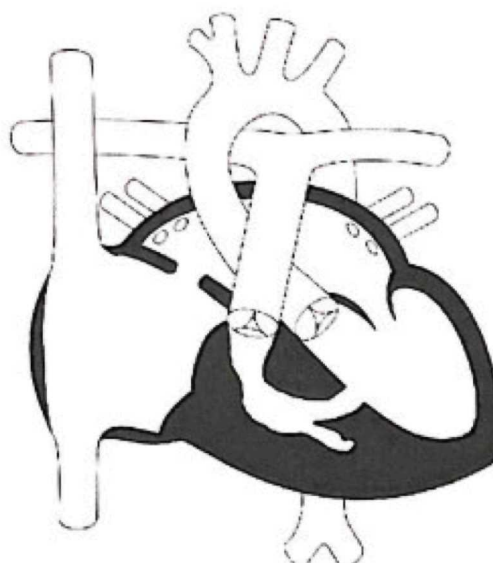
- Heterogeneous group of lesions characterized by complete mixing of systemic and pulmonary venous circulations
- Usually associated with atresia or severe stenosis of a semilunar or atrioventricular valve
- **Hypoplastic Left Heart Syndrome**
 - 1-3% of all congenital heart defects with male predominance
 - Mitral and aortic valve stenosis or atresia associated with a hypoplastic left ventricle
 - Common subtypes: MA/AA, MS/AS, MS/AA (worst prognosis); MA/AS (rare without VSD)
 - High risk lesion: significant portion of neonatal deaths from congenital heart disease
 - Common syndromes
 - Kabuki
 - Noonan
 - Smith-Lemli-Opitz
 - Holt-Oram
 - Ellis-van Creveld
 - Turner and CHARGE syndromes
 - Trisomy 13, 18 and 21
 - Ascending aorta is always hypoplastic, 80% have coarctation
 - 35% have associated tricuspid valve abnormality
 - Severe atrial restriction or intact atrial septum results in rapid deterioration after birth and high mortality risk



Hypoplastic Left Heart Syndrome

- **Unbalanced Atrioventricular Septal Defect (AVSD)**
 - Defect of superior and inferior endocardial cushion fusion during development
 - ~50% of patients with AVSD have Trisomy 21
 - Heterotaxy syndrome is common (right>left isomerism)
 - 10% of AVSDs have unbalanced AV valves committed more to the same ventricle
 - Most have dominant right ventricle with hypoplastic LV
 - Associated with conal defects (DORV or TOF)
 - Double orifice left AV valve increases risk of stenosis
 - Risk of LVOTO due to narrowing from gooseneck deformity and abnormal chordae attachments
 - If severe atrial septal malalignment leftward, risk of pulmonary vein isolation
 - Most common associated defects: secundum ASD, PDA (44%) and LSVC to CS
- **Double Inlet Left Ventricle (DILV)**
 - Two distinct inlets or AV valves to a single ventricle
 - Usually the LV is dominant and the RV is hypoplastic
 - May have D-looped or L-looped ventricles and often have transposed great arteries
 - Holmes Heart: DILV with normally related great arteries.
 - Ventricular communication often termed bulboventricular foramen or outlet foramen.

- Restriction across the bulboventricular foramen can cause progressive LVOT obstruction
- Tricuspid and mitral valve chords may cross to the opposite ventricle and cause outflow tract obstruction
- Coarctation seen with sub-aortic obstruction
- **Tricuspid Atresia**
 - Absent tricuspid valve and no RA-RV communication
 - Obligate ASD or stretched PFO
 - Varying degrees of RV and pulmonary valve hypoplasia related to VSD size
 - Classified by great artery position (I/II) and VSD size (A-C)
 - I = normally related great arteries (70-80%)
 - II = D-transposition
 - A = no VSD and pulmonary atresia
 - B = moderate VSD with some pulmonary stenosis
 - C = large VSD with no pulmonary stenosis
 - Other defects seen in <20%: coarctation (8%), LSVC, right aortic arch



Tricuspid Atresia

Preoperative Physiology

- Physiology varies based upon
 - Whether there is ductal dependency for pulmonary or systemic blood flow
 - Amount of restriction to pulmonary venous return at the atrial septum
- As PVR falls, excessive pulmonary blood flow may result in congestive heart failure, pulmonary edema, and/or shock

- Treatment goal is to maximize systemic oxygen delivery not arterial saturation
- Minimizing oxygen consumption with mechanical ventilation, sedation and muscle relaxation plays an important role in the optimizing Qp/Qs
- Ductal-Dependent Systemic Blood Flow (i.e. HLHS)
 - Prostaglandin infusion necessary to maintain ductal patency and systemic blood flow
 - Undiagnosed neonates often present with circulatory collapse and cardiogenic shock within the first week of life when ductus closes
 - Mild to moderate cyanosis unless a restrictive atrial septum present
 - If atrial septum is unrestrictive, congestive heart failure and low cardiac output can develop as PVR falls and Qp/Qs rises.
 - In presence of severely restrictive atrial septum, profound desaturation and respiratory failure develop shortly after birth
 - Need for urgent atrial septostomy
 - In the setting of adequate oxygen delivery, target arterial saturations are 75-85%
 - As PVR falls, Qp/Qs and saturations will rise
 - Initiate feeds with caution as there is a high risk of necrotizing enterocolitis
 - If signs of impaired oxygen delivery, consider intubation and mechanical ventilation
 - Consider diuretics to manage symptoms of heart failure as Qp/Qs rises
- Ductal-Dependent Pulmonary Blood Flow (i.e. PA/IVS)
 - Cyanosis dependent on size of PDA and additional sources of pulmonary blood flow
 - Prostaglandin infusion to maintain ductal patency
 - In the presence of additional sources of pulmonary blood flow, trial off prostaglandin may be needed to determine necessity of ductus to maintain adequate pulmonary blood flow
 - Target arterial saturation > 75%
 - In balanced circulation without symptoms of CHF, preoperative feeds may be initiated
- Non-Ductal Dependent with Pulmonary Stenosis (i.e. unbalanced AVSD with PS)
 - Degree of cyanosis and amount of pulmonary blood flow (Qp/Qs) are dependent on severity of pulmonary stenosis, size of pulmonary valve annulus, and pulmonary vascular resistance; usually only mild cyanosis

- With minimal PS, signs of congestive heart failure may present early as PVR falls
- Symptoms of heart failure should guide medical management
- Consider pulmonary artery band in patients who fail maximal medical management

Section 2
Pulmonary Artery Band
Molly Lappe MSN, RN, CCRN

Purpose

- Limit pulmonary blood flow

Operative Procedure

- Placed via median sternotomy or left thoracotomy
- Mesh band is tightened around the main pulmonary artery until adequate drop in arterial saturation is seen
- Goal saturation usually 80%
- Judging appropriate tightness of band is an art and readjustment in early postoperative period may be required
- Anchor sutures are used to prevent distal migration of band onto branch pulmonary arteries

Postoperative Considerations and Management

- Significant flow restriction can be present without decrease in SpO₂
- In the absence of significant lung disease, arterial saturations should be used to guide assessment adequacy/tightness of PA band
- Residual signs of pulmonary over-circulation may be present due to loose band, usually in conjunction with high arterial saturations and signs of CHF
- Vasoactive agents rarely required
- Prolonged mechanical ventilation rarely required unless band is too loose and residual signs of congestive heart failure or postoperative inflammatory reactions
- Diuretic duration and requirement dependent on amount of flow restriction from the band

Lesion Specific Postoperative Concerns

- Band migration onto branch pulmonary artery
 - Differential pulmonary blood flow may be seen on CXR
- Band too tight
 - Systemic desaturation
 - Revision required only if desaturation profound with signs of inadequate oxygen delivery
- Band too loose
 - Ongoing signs of pulmonary over-circulation and congestive heart failure
 - Inability to wean from mechanical ventilation
- Increased atrioventricular valve regurgitation from increased afterload

- Subaortic obstruction in patients with bulboventricular foramen or restrictive VSD

Section 3
Blalock Taussig Shunt
Molly Lappe MSN, RN, CCRN

Purpose

- Provide stable source of controlled pulmonary blood flow

Operative Procedures

- Classic Blalock-Taussig shunt (BT shunt): subclavian artery to right pulmonary artery, rarely if ever performed today
- Modified BT Shunt: usually 3-3.5 mm shunt from the systemic artery (typically the right subclavian) to the pulmonary artery
- Central shunt: synthetic tube usually 3-3.5 mm from the aorta to the pulmonary artery
- Typically performed through median sternotomy

Postoperative Considerations and Management

- Continuous diastolic run off through shunt places coronary perfusion at risk due to low diastolic pressure
- Shunt provides fixed resistance, though changes in pulmonary vascular and systemic vascular resistance may lead to changes in Qp/Qs
- Inotropic medications or fluid administration may be required to maintain adequate perfusion pressure of shunt
- Minimize FiO₂ to limit pulmonary vasodilation and rise in Qp/Qs
- Target oxygen saturation 80%
- Sudden drop in saturation and/or ETCO₂ suggest shunt thrombosis and necessitates urgent investigation with auscultation and echocardiography
- Extubation after stable balance of Qp/Qs established
- Differential diagnosis for low arterial saturation: shunt kink or thrombus, pulmonary artery branch stenosis, low cardiac output/hypotension, or lung disease
- Increased risk of necrotizing enterocolitis due to diastolic run off

Lesion Specific Postoperative Concerns

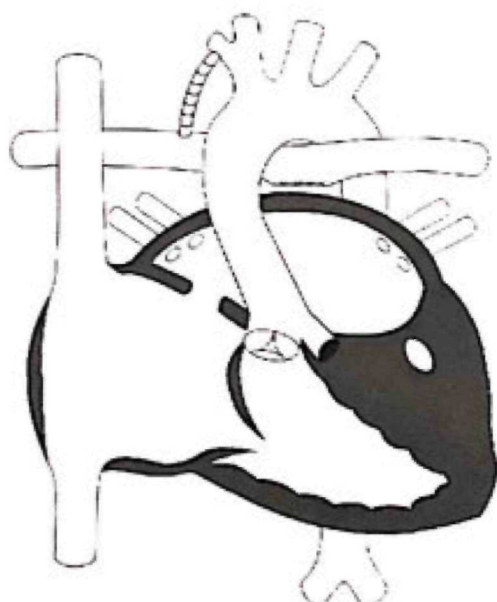
- Shunt thrombosis
 - Sudden decrease in oxygen saturation and ETCO₂
 - Loss or decreased shunt murmur
 - BP initially preserved, eventual hypotension due to poor myocardial oxygen delivery
 - Obstruction can be partial or complete
 - Treatment
 - Heparin bolus (typically 100units/kg)
 - Increase SVR (epinephrine, phenylephrine)
 - Decrease PVR (hyperventilate, increase inspired oxygen)

- Emergent CV surgery or interventional catheterization consult
- Excessive pulmonary blood flow
 - Typically presents with high saturation, persistent tachycardia, congested chest x-ray, failure to wean from mechanical ventilation, and diuretic requirement
 - Management goal is to maintain adequate oxygen delivery and decrease Qp/Qs
 - Decrease SVR
 - Increase PVR (increase PEEP, minimize FiO₂)
 - Decrease metabolic demand
 - Diuretics usually needed

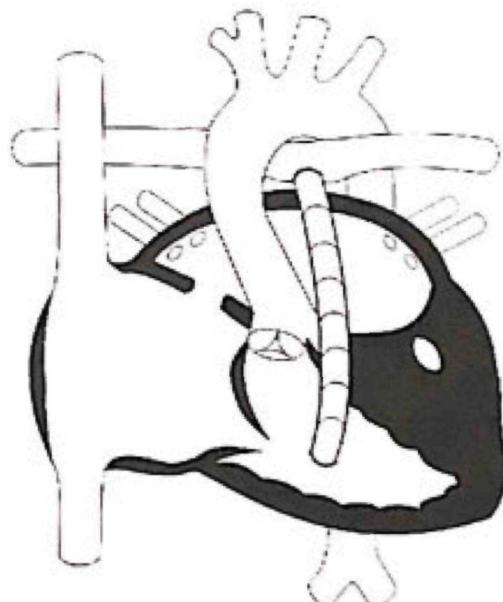
Section 4
Norwood Procedure
Molly Lappe MSN, RN, CCRN

Operative Procedure

- Arch reconstruction using pulmonary artery and homograft material
- Establish a source of pulmonary blood flow with a modified BT shunt or RV to PA conduit (Sano shunt)
- Atrial septectomy
- Performed under deep hypothermic circulatory arrest or low flow CPB
- Sano versus modified BT Shunt
 - Advantages of Sano: lack of continuous diastolic run off and increased diastolic blood pressure
 - Disadvantages of Sano: RV incision in systemic right ventricle, more cyanotic compared to BT shunt with potential need for earlier BDG



Norwood-BT Shunt



Norwood-Sano

Postoperative Consideration and Management

- Low diastolic blood pressure from run off through a modified BT shunt can compromise coronary perfusion
- Patients with Sano shunt usually more cyanotic than those with modified BT shunt
- As myocardial function recovers and cardiac output increases, arterial saturation may increase and does not necessarily herald excessive pulmonary blood flow
- Arrhythmias are concerning, monitor closely for signs of adequate oxygen delivery

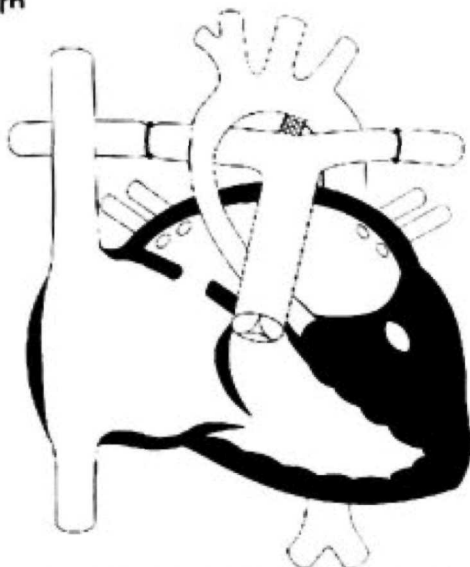
Lesion Specific Postoperative Concerns

- Persistent low cardiac output syndrome (refer to Chapter 3 Section 2)
 - Transient decrease in systemic perfusion resulting in an imbalance between oxygen delivery and oxygen consumption
 - Typically occurs between 6-18 hours after surgery
 - Anticipation of LCOS can aid in diagnosis and prompt treatment
 - Consider evaluation (echo and/or catheterization) to evaluate aortic arch, coronary arteries, AV valves, and ventricular function
- Arch obstruction
 - Signs of low cardiac output or worsening myocardial function even in presence of mild echocardiographic gradients
 - Blood pressure differential between upper and lower extremities (depending on the location of the obstruction)
 - Feeding intolerance
 - Diminished distal pulses
- Shunt thrombosis
 - Sudden decrease in oxygen saturation and ETCO_2
 - Loss or decreased shunt murmur
 - BP initially preserved, eventual hypotension due to poor myocardial oxygen delivery
 - Obstruction can be partial or complete
 - Treatment
 - Heparin bolus (typically 100units/kg)
 - Increase SVR (epinephrine, phenylephrine)
 - Decrease PVR (hyperventilate, increase inspired oxygen)
 - Emergent CV surgery or interventional catheterization consult
- Excessive pulmonary blood flow
 - Typically presents with high saturation, persistent tachycardia, congested chest x-ray, failure to wean from mechanical ventilation, and diuretic requirement
 - Management goal is to maintain adequate oxygen delivery and decrease Qp/Qs
 - Decrease SVR
 - Increase PVR (increase PEEP, minimize FiO_2)
 - Decrease metabolic demand
 - Diuretics usually needed

Section 5
Hybrid Procedure
Afrothite Kotsakis, MD, Med, FRCPC

Operative and Interventional Procedure

- Combination of interventional catheterization and pulmonary artery bands to substitute for the Norwood procedure
- Bilateral PA bands placed on each branch PA to prevent excessive pulmonary blood flow
- PDA stent placed to maintain systemic blood flow without the need for prostaglandin
- Balloon atrial septostomy performed to assure unobstructed pulmonary venous return



HLHS with Hybrid palliation (Bilateral PA bands, PDA stent)

Postoperative Consideration and Management

- Post-procedure physiology similar to preoperative physiology
- Patients continue to have single ventricle parallel circulation with low diastolic blood pressure
- Goal is to optimize systemic oxygen delivery
- Vasoactive agents may be required
- Close monitoring of adequacy of oxygen delivery recommended
- Ventilation management goals include avoiding atelectasis, alkalosis, and excessive hypoxia
- If supplemental oxygen required, target SaO₂ 75-85%
- Patients with single ventricle physiology are at increased risk for necrotizing enterocolitis compared to other postoperative neonates
 - Use of standardized feeding protocol associated with reduced incidence

- Sedation to reduce metabolic demand useful when oxygen delivery is impaired

Lesion Specific Postoperative Concerns

- Inadequate pulmonary artery banding
 - Signs include low cardiac output and pulmonary edema
 - May be unilateral with one lung overperfused and one lung underperfused
- PDA obstruction
 - Etiologies
 - Migration of the stent
 - Uncovered portions of the PDA
 - Retrograde obstruction by stent struts can occur leading to coronary and cerebral artery insufficiency
 - Signs include low cardiac output and myocardial dysfunction
- Restrictive atrial septum
 - Signs include pulmonary edema, cyanosis
 - May require stent across atrial septum

CHAPTER 10

Cavopulmonary Connections

Chapter Editors

Harris Baden, MD

Nancy Rudd, MS, APNP, CPNP-PC/AC

Section 1: Superior Cavopulmonary Connection

Section 2: Fontan

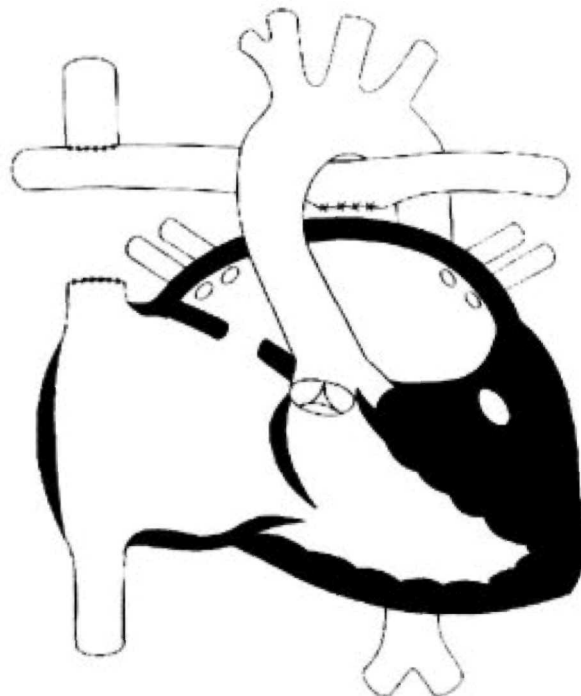
Section 1
Superior Cavopulmonary Connection
Katherine Banker, MD and Shelina M. Jamal, MD, FRCPC

Preoperative Evaluation

- Preoperative evaluation traditionally includes echocardiogram and cardiac catheterization
- Some patients may undergo cardiac computerized tomography (CT scan) or magnetic resonance imaging (MRI) and catheterization is deferred
- Ideal preoperative hemodynamics
 - Low pulmonary artery pressure (< 15 mmHg) and pulmonary vascular resistance (PVR) < 4 Wood units
 - Unobstructed pulmonary venous return
 - Minimal AV valve regurgitation
 - Normal ventricular function and common atrial pressure (< 10 mmHg)
 - Unobstructed systemic blood flow

Operative Procedure

- Two options for performing superior cavopulmonary connection (SCPC)
 - Bidirectional Glenn operation
 - An end-to-side anastomosis of the SVC to branch PA
 - SVC is divided at level of branch PA, the cardiac side is oversewn and cranial side is anastomosed to the superior aspect of the branch PA
 - Provides blood supply to both PAs (bidirectional)
 - Can be performed with or without cardiopulmonary bypass
 - Hemi-Fontan Operation



Bidirectional Glenn

- A confluence of the cardiac end of the SVC, the RA and the RPA is created. A dam of homograft tissue across the superior cavoatrial junction separates SVC blood flow from IVC and coronary sinus blood flow
- Both operations typically involve closure of systemic to PA shunt and any additional sources of pulmonary blood flow (i.e., oversewing of native pulmonary valve)

Postoperative Consideration and Management

- Pulmonary blood flow is passive and anatomic or physiologic obstruction at any level will compromise blood flow
- Optimal cerebral blood flow (CBF)
 - $\uparrow pCO_2 = \uparrow CBF = \uparrow SCPC \text{ flow}$
 - $\downarrow pCO_2 = \downarrow CBF = \downarrow SCPC \text{ flow}$
- Avoiding lung overdistension and atelectasis optimizes PVR and SCPC flow
- Acute volume unloading of the single ventricle leads to ventricular volume to muscle mass mismatch and diastolic dysfunction; may require volume to achieve a higher common atrial pressure
- Often hypertensive, requiring vasodilators
- If mechanically ventilated, minimize airway pressure and attempt extubation as soon as possible, as spontaneous ventilation enhances pulmonary blood flow through the SCPC
- Passive SCPC flow increases risk of thrombosis: must remove SCPC lines as soon as possible and anticoagulate per institutional practice

Lesion Specific Postoperative Concerns

- Hypertension
 - Causes
 - Neurohormonal activation and increase in SVR
 - Pain and anxiety
 - Acute increase in SVC pressure leads to $\uparrow$ cerebral venous pressure, leading to compensatory $\uparrow$ systemic BP to maintain cerebral perfusion pressure
 - Acute volume unloading of systemic ventricle
 - Treatment
 - Sedation and analgesia
 - Elevate head, midline
 - Often require vasodilators for acute management
- Hemodynamic Profiles
 - High transpulmonary gradient (i.e., high CVP and low common atrial pressure)
 - High PVR: lung parenchymal disease, high airway pressures, pleural effusion
 - Obstruction/thrombus at surgical anastomosis, pulmonary artery, or pulmonary vein

- May present as SVC syndrome, venous congestion associated with swelling of the head
- High common atrial pressure and high CVP with normal transpulmonary gradient
 - AV valve stenosis or regurgitation
 - AV dissociation
 - Ventricular dysfunction, most often diastolic dysfunction
 - Outflow obstruction
 - Pericardial effusion/tamponade
- Low common atrial and CVP pressures
 - Inadequate preload
- Impaired oxygenation
 - Causes
 - Inadequate pulmonary blood flow: evaluate for anatomic obstruction to flow and elevated PVR
 - Pulmonary venous desaturation: evaluate for parenchymal or pleural disease
 - Systemic venous desaturation: evaluate for low cardiac output, anemia, or any condition increasing oxygen consumption (i.e., fever, agitation)
 - Decompressing venous collaterals (SVC decompressing to IVC)
 - Treatment
 - Permissive hypercapnia may be used to improve cerebral perfusion, SCPC flow and oxygenation in those with inadequate pulmonary blood flow
 - While unlikely, PVR may be elevated postoperatively and a trial of iNO may be indicated
 - Optimize ventricular function and reduce afterload
 - Transfuse for anemia and minimize oxygen consumption
 - Optimize lung volume, evacuate effusions, and pneumothoraces

Section 2

Fontan

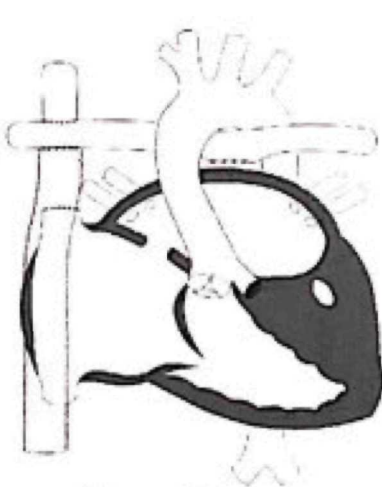
Titus Chan, MD, MS, MPP and Monique Radman, MD, MAS

Preoperative Evaluation

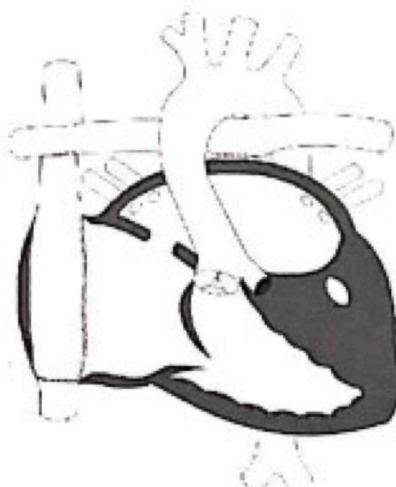
- Candidates for a Fontan operation have typically undergone a superior cavopulmonary connection (Bidirectional Glenn or Hemi-Fontan) or a hemiazygous to pulmonary artery anastomosis procedure (Kawashima)
- Ideal preoperative hemodynamics
 - PVR < 4 Wood units
 - Mean pulmonary arterial pressure < 15 mm Hg
 - Good systemic ventricular function (cardiac index > 3.0 L/min/m²; EDP < 10 mm Hg)
 - AV valve competency
 - No pulmonary artery obstruction
 - Sinus rhythm

Operative Procedures

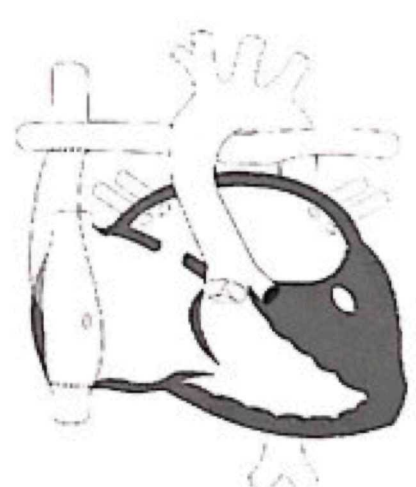
- Inferior vena cava (IVC) is redirected to the pulmonary artery (PA)
 - Extracardiac Fontan: conduit external to the common atrium connects the IVC to the PA
 - Lateral tunnel Fontan: intra-atrial baffle within the common atrium connects the IVC and main PA
 - Intra-extracardiac Fontan: ring supported conduit sutured to internal orifice of IVC. Conduit emerges from side of RA and is sutured to Glenn anastomosis
- A fenestration may be placed between the Fontan circuit and the common atrium to allow for a right to left decompression and augmentation of ventricular preload



Extracardiac Fontan



Lateral Tunnel Fontan



Intra-Extracardiac Fontan

Postoperative Considerations and Management

- Results in completely passive venous blood flow to the PAs
- Goal hemodynamics: common atrium: 5-10 mm Hg; Fontan/CVP: 10-15mmHg; transpulmonary gradient <10mmHg (Fontan/CVP - common atrial pressure)
- Systolic dysfunction is uncommon; varying degrees of diastolic dysfunction may be responsive to volume administration
- AV dissociation generally is poorly tolerated
- Optimize ventilation and oxygenation to minimize PVR while avoiding hyperventilation so as not to compromise SVC flow
- Early extubation is associated with increased pulmonary blood flow and cardiac output
- Aggressively evacuate pleural effusions and pneumothoraces

Lesion Specific Postoperative Concerns

- Hemodynamic profiles
 - High transpulmonary gradient (i.e., high CVP and low common atrial pressure)
 - High PVR: lung parenchymal disease, high airway pressures, pleural effusion
 - Obstruction/thrombus at surgical anastomosis, pulmonary artery, or pulmonary vein
 - High common atrial pressure and high CVP with normal transpulmonary gradient
 - AV valve stenosis or regurgitation
 - AV dissociation
 - Ventricular dysfunction, most often diastolic dysfunction
 - Outflow obstruction
 - Pericardial effusion/tamponade
 - Low common atrial and CVP pressures
 - Inadequate preload
- Impaired oxygenation
 - Veno-venous collaterals
 - Excessive right to left shunting at a fenestration
 - Pulmonary venous desaturation
- Ascites
 - May require peritoneal drain
- Prolonged pleural effusions/chylothorax
 - Loss of protein and immunoglobulins
- Hypercoagulable state
 - Secondary to hepatic dysfunction and protein loss
 - High risk of venous thrombosis
 - Anticoagulation per institutional practice

CHAPTER 11

Miscellaneous Lesions

Chapter Editors

Roxanne Kirsch, MD, FRCPC, FAAP
Milagros Tablante, MSN, ARNP, PNP-BC

Section 1: Anomalous Left Coronary from Pulmonary Artery

Section 2: Anomalous Aortic Origin of Coronary Artery

Section 3: Vascular Rings and Slings

Section 4: Pulmonary Vein Stenosis

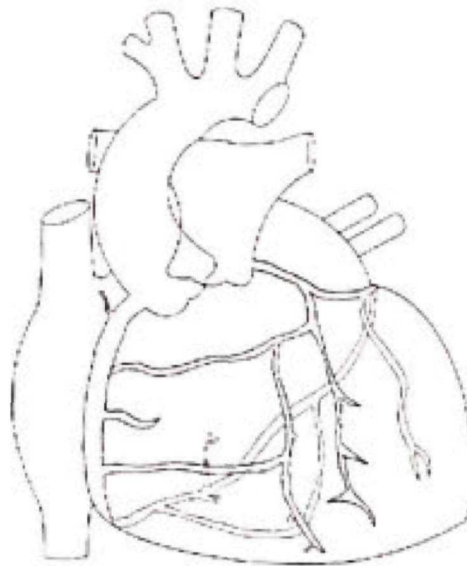
Section 5: Cor Triatriatum

Section 6: Double Outlet Right Ventricle

Section 1
Anomalous Left Coronary from Pulmonary Artery
Michael Hobson, MD and Trina Blunk, MSN, APRN, CPNP-PC

Anatomy

- Left coronary artery (LCA) arises from the pulmonary artery instead of the aorta
- Right coronary artery (RCA) from the main pulmonary artery (MPA) or both coronaries from the MPA have been described
- Diagnosis is difficult, 90% mortality in the first year of life if left untreated



ALCAPA

Preoperative Physiology

- In the early neonatal period, LCA is well perfused due to elevated PVR and PA pressure near systemic, exceeding intramural LV pressure
- As PVR decreases and the ductus closes, LCA receives desaturated blood at lower pressure resulting in progressive LV ischemia and dilation
- Progressive LV dilation and/or papillary muscle ischemia leads to mitral regurgitation
- Collateral vessels from RCA to LCA may form with flow reversal from LCA to PA. Left to right shunting (RCA-LCA-PA) exacerbates ischemia (coronary steal)
- High suspicion in any patient presenting with mitral insufficiency and LV dilation in the first few months of life. May progress to dilated cardiomyopathy

- Depending on how collaterals develop, patients may be asymptomatic, but later in life may experience arrhythmias, angina, or sudden death
- ECG findings of Q waves in Lead I and aVL without Q waves in II, III, aVF are highly suggestive of ALCAPA
- Echocardiogram provides diagnosis in most cases
 - RCA dilation (due to increased flow) and retrograde LCA flow with diastolic jet into the MPA, ischemic changes of papillary muscles and LV endocardium
- Cardiac catheterization should be performed if echocardiogram images are insufficient but clinical suspicion for ALCAPA remains high
- Medical management is aimed at stabilizing patient prior to urgent surgical repair

Operative Procedures

- Surgical intervention should be performed as early as possible to reduce ongoing ischemia
- Goal of repair is to establish a two coronary system
- Type of surgical repair based on origin of the coronary artery
- Mitral valve usually not addressed initially due to likelihood of improvement as LV function improves
- Two common techniques for repair
 - Reimplantation of the anomalous left coronary artery into the aorta—preferred method
 - Intrapulmonary baffle (Takeuchi procedure)
 - Aortopulmonary window created
 - Baffle created utilizing the anterior wall of the pulmonary artery, synthetic material, or cryopreserved homograft and placed from aorta to the orifice of the anomalous coronary ostium within the PA
 - Augmentation of the anterior pulmonary artery may be required so as not to impede PA flow from coronary baffle on posterior wall of the PA

Postoperative Consideration and Management

- LV dysfunction will persist in the immediate postoperative period from cardiopulmonary bypass and LV reperfusion injury
- Initially requires inotropic support and aggressive afterload reduction due to ventricular dysfunction and mitral insufficiency
- Long-term afterload reduction may be needed (reduces LV wall stress and decreases MR)
- Avoid overly aggressive fluid resuscitation or excessive catecholamines
- ECMO or LVAD may be required in cases with severely depressed ventricular function
- Usually remains on mechanical ventilatory support for 24-48 hours, longer support may be needed to support LV dysfunction

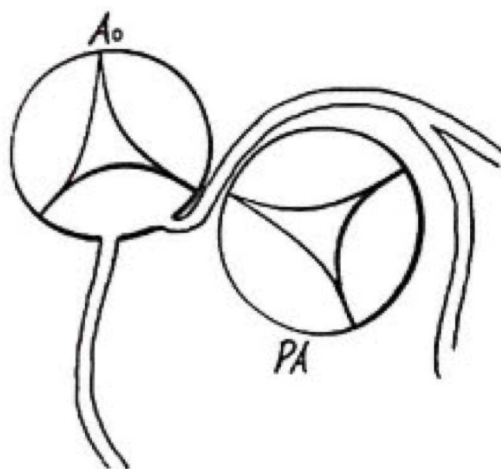
Lesion Specific Postoperative Concerns

- Ventricular dysrhythmias
 - Due to LV injury/ischemia, rule out obstructed LCA flow postoperatively
- Low cardiac output, LV dysfunction, and mitral insufficiency
 - Gradual improvement in ventricular function over months
 - Myocardial recovery often seen within first year postoperatively
 - Mitral regurgitation takes longer to resolve than LV function and may not completely resolve in all cases
 - May require reoperation for mitral valvuloplasty or mitral valve replacement later in life
- Coronary spasm
 - Risk due to surgical manipulation of coronary artery
 - Nitroglycerin infusion may be utilized for coronary perfusion

Section 2
Anomalous Aortic Origin of the Coronary Artery
Michael Hobson, MD and Trina Blunk, MSN, APRN, CPNP-PC

Anatomy

- Anomalous origin of the left coronary artery from the right sinus of Valsalva
 - Right coronary ostium is at its normal location (center of the right sinus)
 - Left coronary ostium arises from the right sinus and is often slit-like and stenotic
 - LCA courses posteriorly between the aorta and the main pulmonary artery (interarterial coronary artery) and courses within the muscle (intramural)
 - Reaches normal epicardial course, opposite to the left sinus of Valsalva
 - Association with TGA



Anomalous Aortic Origin of the Coronary Artery - LCA from right

- Anomalous origin of the right coronary artery from the left sinus of Valsalva
 - Right coronary ostium arises from the left sinus of Valsalva
 - Courses anteriorly between the great arteries and is often intramural and stenotic
 - Reaches normal epicardial course, opposite to the right coronary sinus.
- Rare high risk anomalies

- Anomalous left or right coronary artery arising from the wrong sinus and coursing intramural in the subpulmonary muscular conus
- Anomalous left anterior descending artery arising from the right sinus and coursing between the great arteries

Preoperative Physiology

- High risk of sudden cardiac death during or just after exercise
- Risk of sudden cardiac death is increased when interarterial course between the great arteries has intramural segment, is hypoplastic, stenotic, or has an abnormal orifice
- Coronary arteries that course between the aorta and pulmonary artery are thought to be at risk for intermittent compression, especially noted with exercise
- Diagnosed with echocardiogram, cardiac MRI, or catheterization with coronary angiography
- Usually asymptomatic, however will occasionally present with signs of intermittent or ongoing myocardial ischemia, including syncope, angina, and congestive heart failure

Operative Procedures

- No medical therapy is available
- Two different surgical techniques
 - Direct repair of the anomalous origin in the aortic root
 - Unroofing the entire coronary artery or creating a new ostium at the distal end of the ostium
 - In the case where left and right coronary arteries' orifices are separated but in the same sinus, the anomalous coronary is reimplanted to the correct aortic sinus
 - When long segments are stenotic, a saphenous vein patch may be utilized to enlarge artery prior to reimplantation
 - Coronary artery bypass grafting
 - Generally avoided in young patients due to limited longevity of the vein grafts

Postoperative Consideration and Management

- May need inotropic support for 24-48 hours, depending on degree of preoperative LV dysfunction
- Monitor for signs of cardiac ischemia after repair including ST segment elevation or depression, arrhythmia, and ventricular dysfunction
- Screening tests include CK-MB profile, CPK, and troponin
- Monitor routine echocardiogram to evaluate for unobstructed coronary ostia
- If requiring higher than expected amount of sedation, may consider obtaining 12-lead ECG to rule out coronary ischemia

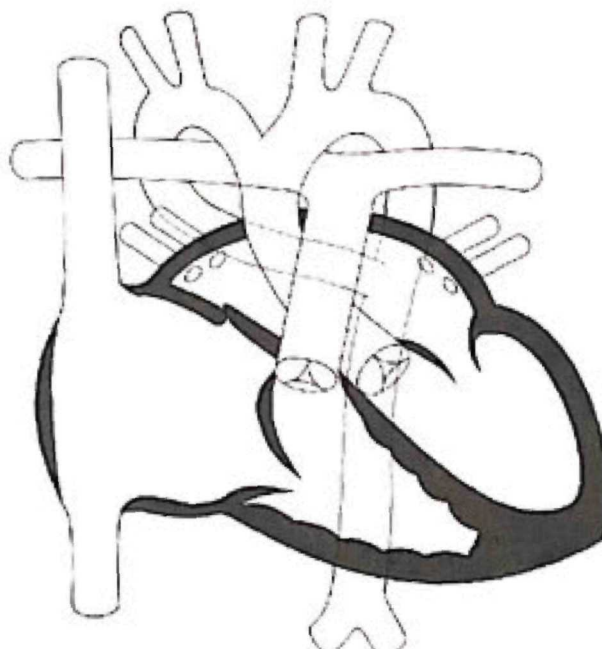
Lesion Specific Postoperative Concerns

- Cardiac ischemia
 - Risk of kinking or tension of coronary artery following coronary artery bypass grafting resulting in cardiac ischemia
 - Echocardiographic findings of regional wall motion abnormalities in the affected coronary region indicate cardiac ischemia
 - May require reoperation, inotropic support.
 - Risk of coronary spasm due to coronary manipulation
 - Nitroglycerin infusion may be utilized for coronary vasodilation

Section 3
Vascular Rings and Slings
Jana Norris, MSN, CNS, CNPNP

Anatomy: Complete Vascular Rings

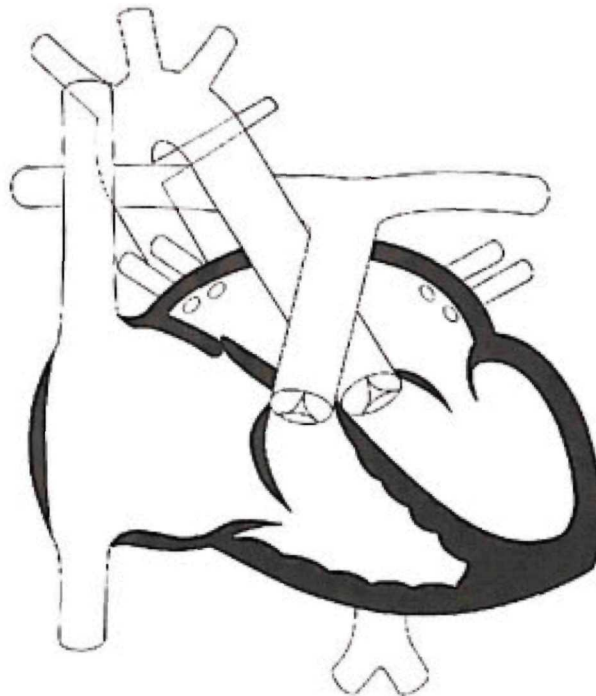
- Vascular rings are vascular anomalies that occur as a result of abnormal development of the aortic arch and cause compression of the trachea, esophagus, or both
- Double aortic arch (most common)
 - Embryologic right and left fourth arches persist causing double arch to form
 - A complete ring is formed when the aortic arches arising from the ascending aorta and passes on both sides of the trachea and esophagus to join at the descending aorta forming a ring around the esophagus and trachea



Double Aortic Arch

- Right aortic arch
 - Multiple configurations of a right aortic arch depending on site of left arch interruption and branching pattern to the left subclavian artery, left carotid artery, and ductus arteriosus
 - Two most common configurations
 - Right aortic arch with retroesophageal left subclavian artery (most common).

- The aberrant left subclavian artery (LSCA) comes off the descending aorta and courses to the left, going behind the esophagus.
- A complete ring is formed when the ligamentum arteriosum comes off the descending aorta and attaches to the left pulmonary artery (LPA)



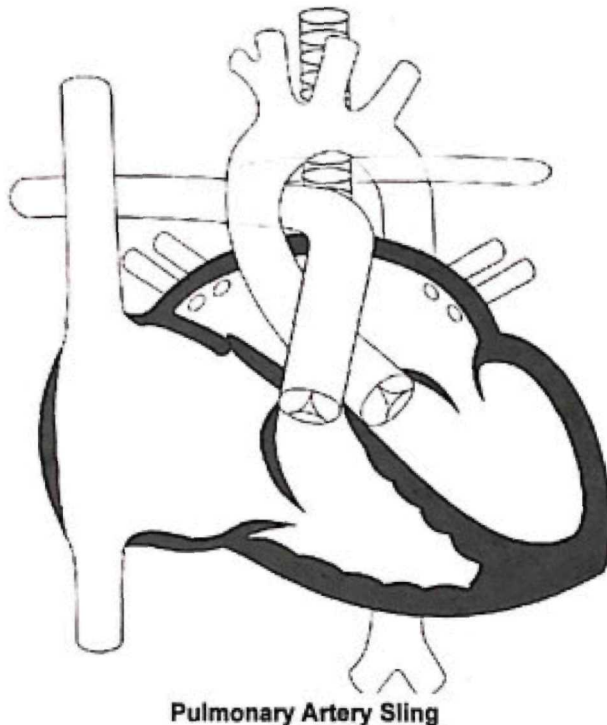
Right Aortic Arch with Anomalous Origin of the Left Subclavian Artery

- Right aortic arch with mirror image branching and left ligamentum arteriosum.
- A complete ring is formed when the ligamentum arteriosum comes off the descending aorta

Anatomy: Partial Vascular Rings

- Innominate artery compression syndrome
 - Anterior tracheal compression caused by an innominate artery that originates more posteriorly and leftward from the aorta
- Left pulmonary artery (LPA) Sling
 - LPA originates from the RPA rather than the main pulmonary artery
 - Tracheal compression occurs when the LPA courses around the right main bronchus and between the trachea and esophagus forming a sling

- Commonly associated with complete tracheal rings



- Aberrant right subclavian artery
 - Esophageal compression caused by presence of right subclavian artery that comes off descending left aorta and courses rightward behind the esophagus

Preoperative Physiology

- Narrowing of trachea and/or esophagus depending on type
- Symptoms of increased work of breathing: retractions, flaring, and tachypnea
- Infants may hyperextend their neck (to decrease amount of tracheal narrowing/compression)
- History of noisy breathing, stridor, or frequent upper respiratory tract infections.
- History of gastroesophageal reflux, feeding/swallowing difficulties, or choking
- Chest radiograph can demonstrate right arch
- Barium esophogram will show indentation of the esophagus
- Bronchoscopy can confirm compression but not diagnose anomaly and diagnose complete tracheal ring

- Echocardiography may identify aberrant vasculature and associated anomalies
- CT or MRI provide the best imaging of both the vasculature and the airway

Operative Procedures

- Double aortic arch
 - Division of lesser of two arches, ligate and divide ligamentum arteriosum via left thoracotomy
- Right aortic arch with left ligamentum arteriosum
 - Ligate and divide ligamentum, resect Kommerell's diverticulum (if present), and transfer left subclavian artery to left carotid via left thoracotomy
- Innominate artery compression syndrome
 - Suspend innominate artery from posterior sternum after thymus resection via right thoracotomy
- PA sling
 - On cardiopulmonary bypass, ligate and divide LPA, implant LPA into MPA anterior to trachea via median sternotomy
 - If complete tracheal rings present, slide tracheoplasty or tracheal reconstruction performed

Postoperative Considerations and Management

- Generally do not require vasoactive infusions unless vascular ring associated with another anomaly
- Tracheal reconstruction for complete tracheal rings may require prolonged intubation, otherwise usually extubated within 12-24 hours
- Often continue to have oxygen requirement in postoperative period
- Should not require diuretics unless cardiopulmonary bypass needed
- Monitor for development of chylous effusions with initiation of food products containing long chain triglycerides, if develops may require medium chain triglyceride based formula or low/no fat diet

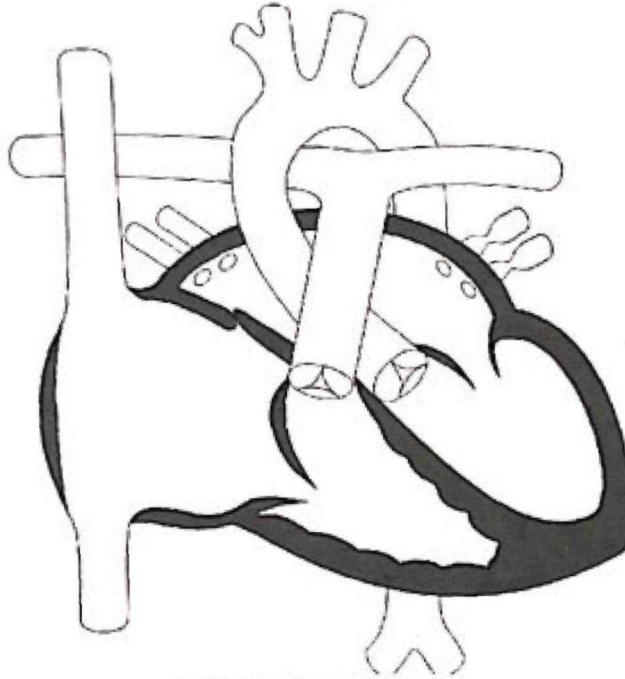
Lesion Specific Postoperative Concerns

- Eventual resolution of symptoms and minimal complications.
- Stridor or increased work of breathing may persist secondary to tracheomalacia.
- Potential for diaphragm or vocal cord paralysis due to proximity of surgical site to phrenic and laryngeal nerve
- Potential for thoracic duct injury and chylous effusions
- Chest tube often not removed until after ingestion of fats

Section 4
Pulmonary Vein Stenosis
Jason Patregnani, MD

Anatomy

- Congenital
 - Fibrous, intimal hyperplasia most commonly affecting the junction where the pulmonary veins connect to the left atrium
 - Progressive over time in almost all cases
 - Can lead to complete obliteration of the vein
 - Associated with a high mortality rate



Pulmonary Vein Stenosis

- Secondary
 - Occurs in up to 10% of patients who have had repair of TAPVR
 - Associated with restrictive or intact atrial septum in hypoplastic left heart syndrome
 - Commonly develops in patients with chronic lung disease

Preoperative Physiology

- Patient will often present with failure to thrive, respiratory distress and hemoptysis with increasing symptoms of pulmonary hypertension over time
- Often misdiagnosed as asthma or chronic lung disease
- Pulmonary venous hypertension is the primary physiologic manifestation
 - Pulmonary edema

- Increased right ventricular pressure leading to right ventricular hypertrophy
- Medical management is palliative
 - Diuresis for the pulmonary edema
 - Sildenafil and/or bosentan are often used, but can worsen the pulmonary edema
 - Chemotherapeutic agents such as methotrexate and imatinib have shown disappointing results in controlling progression of the disease, but trials are ongoing

Operative and Interventional Procedures

- Interventional
 - Balloon and stent angioplasty can be effective in the short term
 - Recurrence of disease in the vast majority of cases with recurrent balloon dilations required for palliation
- Surgical
 - Decompression of the obstruction with resection of tissue and trocar dilation
 - Stenosis almost always recurs
 - Sutureless repair
 - Longitudinal incision of the base of the pulmonary veins, detachment of the left atrium, and reanastomosis of the atrium to the pericardium
 - Recurrence remains common and not as useful if bilateral disease

Postoperative Consideration and Management

- Pulmonary hypertension
- Pulmonary edema
- Milrinone is used for right ventricular relaxation and pulmonary vasodilation
- Duration of vasoactive support dependent on preoperative duration of the disease, the severity of lung disease and pulmonary noncompliance, and severity of pulmonary hypertension
- If long standing disease, pulmonary hypertension may be more problematic, and need longer period of mechanical ventilation
 - Goal pH 7.4-7.45 to avoid elevation of pulmonary vascular resistance
 - May need iNO
 - 100% FiO₂ may be required, although weaning oxygen as patient allows is important to reduce parenchymal injury and oxygen free radicals
- Early diuretics due to long standing pulmonary edema
- Wean sedation slowly at time of extubation, if clinically improving without signs of pulmonary hypertension exacerbation

- Neuromuscular blockade may be warranted if displaying symptoms of pulmonary vascular reactivity

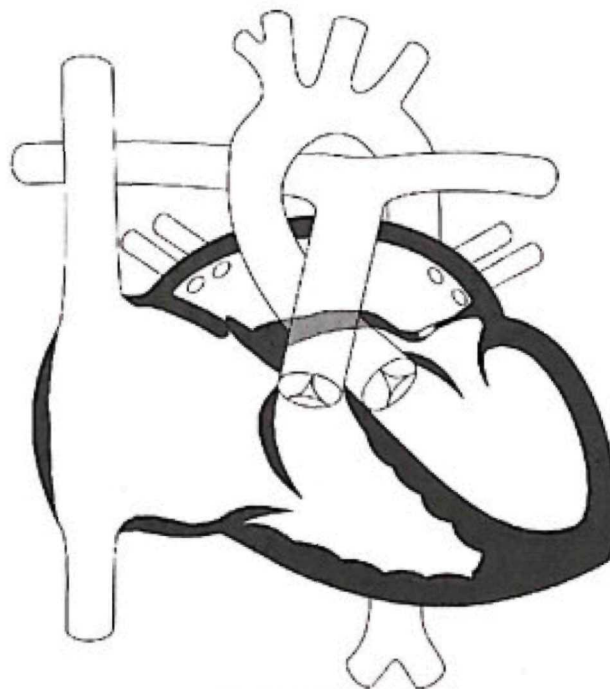
Lesion Specific Postoperative Concerns

- Pulmonary hypertension
 - Pulmonary vasculature can be hyper-reactive in the postoperative period and patient may be at risk for sudden life-threatening pulmonary hypertensive crises
 - Presence of an atrial septal defect is important to note – allows decompression of failing right ventricle but can lead to excessive cyanosis which may not be tolerated
- Right ventricular hypertrophy
 - Decreased right ventricular relaxation
- Residual pulmonary venous obstruction
 - Must be considered in setting of increased RV pressure non-responsive to therapy

Section 5
Cor Triatriatum
Lindsey Ryerson, MD, FRCPC

Anatomy

- Fibromuscular membrane that divides the left atrium into a proximal chamber that receives the pulmonary venous return and a distal chamber that communicates with the left atrial appendage and the mitral valve
- There is a variable sized orifice in the membrane that connects the proximal and distal chambers of the left atrium
- An atrial communication typically connects the right atrium with the distal left atrial chamber
- Common associated cardiac anomalies include patent foramen ovale, secundum atrial septal defect, patent ductus arteriosus, partial or total anomalous pulmonary venous return, left superior vena cava, and pulmonary vein stenosis



Cor Triatriatum

Preoperative Physiology

- Similar physiology to mitral stenosis
- Physiology depends on the degree of intra-atrial obstruction (size of the opening in the membrane connecting the proximal and distal chambers) and the presence or absence of intra-atrial communication
- If the membrane opening is small may develop

- Pulmonary venous obstruction with pulmonary edema
- Pulmonary venous and subsequent pulmonary arterial hypertension
- Low cardiac output secondary to right heart failure

Operative Procedure

- Membrane removed via right or left atrial approach depending on surgical preference and co-existing cardiac disease

Postoperative Consideration and Management

- Pulmonary hypertension will not resolve immediately after membrane resection
- Depending on degree of pulmonary hypertension, RV may need inotropic support immediately after weaning from CPB
- Depending on the age of the patient and degree of pulmonary hypertension, children should be able to extubate within the first 24 hours
- Severe pulmonary hypertension may require prolonged mechanical ventilation

Lesion Specific Postoperative Concerns

- Pulmonary hypertension
 - Pulmonary vasculature can be hyper-reactive in the postoperative period and patient may be at risk for sudden life-threatening pulmonary hypertensive crises
 - Presence of an atrial septal defect is important to note – allows decompression of failing right ventricle but can lead to excessive cyanosis which may not be well tolerated
- Arrhythmias
 - Atrial arrhythmias (i.e., atrial fibrillation or flutter) secondary to left atrial dilation or suture lines

Section 6

Double Outlet Right Ventricle

Dipankar Gupta, MD, Joseph Philip, MD, Mark S Bleiweis, MD, F Jay Fricker, MD
and Jai P Udassi, MD, FACC, FAHA

Anatomy

- Double outlet right ventricle (DORV) indicates the presence of a ventricular-arterial malalignment, not a specific type of congenital heart disease and is heterogeneous group of lesions with variable physiology
- Aorta and pulmonary trunk arise predominantly (> 50% or more) from the RV with failure of aortic conus to regress causing aorta to be more anterior rather than posterior and inferior
- VSD is described in terms of its relation to the great arteries: subaortic, subpulmonary, doubly committed, or non-committed
- Discontinuity of mitral and aortic valves due to subaortic conus is a defining feature of DORV
- DORV with subaortic or doubly committed VSD with PS
 - TOF-type VSD is located just below the aorta
 - Most common type, occurs in 40% of all DORV defects
- DORV with subpulmonary VSD (Taussig-Bing)
 - Transposition-type: bilateral conus, side by side semilunar valves, and subpulmonary VSD lying anteriorly and superiorly beneath the pulmonary valve
 - Occurs in up to 20% of all DORV defects
 - Commonly associated with coarctation of aorta
 - Subaortic stenosis uncommon but significant and usually due to hypoplastic LVOT
- DORV with subaortic VSD without PS
 - VSD type: entire pulmonary trunk and at least half of aorta arise from the RV with VSD most closely aligned with the aorta
- DORV with doubly committed VSD
 - Presence of a large conoventricular VSD below both the aortic and pulmonary valves
- DORV with non-committed VSD
 - VSD is remote from both great arteries, is located within the inlet septum without perimembranous extension
 - VSD may be an inlet (AV canal type)

Preoperative Physiology

- Diverse clinical presentation based on anatomy
- DORV with subaortic or doubly committed VSD with PS
 - Presents with mild to severe cyanosis with gradual worsening of cyanosis, and potentially hypercyanotic spells

- DORV with subpulmonary VSD (Taussig-Bing)
 - Newborns may present cyanotic depending on adequacy of atrial communication and ductus arteriosus
 - May need atrial septostomy prior to surgery
- DORV with subaortic VSD without PS and DORV with doubly committed VSD
 - Presents with left to right shunt causing symptoms of congestive heart failure, including: failure to thrive, pulmonary edema and cardiomegaly
 - Symptoms worsen as PVR decreases.
- DORV with non-committed VSD
 - Presentation dependent on severity of pulmonary stenosis

Operative Procedures

- VSD (position, size, and number), AV valve abnormalities, and associated defects determine the primary surgical procedure and may preclude biventricular palliation
- Inadequate RV size or abnormal AV valves warrants either single ventricle staging or 1 ½ ventricle palliation (biventricular septation plus bidirectional Glenn)
- VSD type and relation to great arteries determines eligibility for biventricular or single stage repair
- Repair delayed with palliation may be performed for symptom relief (i.e, BT shunt for inadequate pulmonary blood flow) in cases with more complex anatomy for possible biventricular repair with growth
- DORV with subaortic or doubly committed VSD with PS
 - Generally repair done by 2-4 months of age
 - Intraventricular tunnel is recreated, connecting the LV to the aorta (VSD is closed with tunnel technique)
- DORV with subpulmonary VSD (Taussig-Bing)
 - Generally repaired before 2 months of age
 - Arterial switch operation (ASO) with tunnel closure of the VSD
 - REV (Reparation l'Etage Ventriculaire)
 - Performed in patients with abnormal ventriculoarterial connections, PS, and low PA pressures
 - Involves technique similar to ASO with right ventriculotomy and the posterior margin of the PA is sutured to the RV and a pericardial patch is used to close the inferior portion of the RV and anterior portion of the PA
 - Nikaidoh
 - Performed in patients with significant PS
 - Involves aortic translocation and biventricular outflow tract reconstruction.
 - Yasui operation
 - Damus-Kaye-Stansel (DKS) with arch reconstruction if needed

- Baffling of VSD to encompass both semilunar valves
- Placement of an RV to PA conduit or single ventricle staging
- BT shunt may be indicated in patients with anomalous coronary crossing the RVOT or if severe branch PA stenosis is present
- DORV with subaortic VSD without PS
 - An intraventricular tunnel repair connecting the LV to the aorta
- DORV with doubly committed VSD
 - Repair in early infancy due to risk of development of pulmonary vascular disease
 - Creation of intraventricular tunnel connecting the LV to the aorta
- DORV with non-committed VSD
 - May require systemic to pulmonary artery shunt as newborn depending on amount of PS
 - May require single ventricle palliation
 - May require REV if it is necessary to place PA on the LV side of the patch
 - May require enlargement of VSD and RVOT muscle bundle resection

Postoperative Considerations and Management

- Patients with excessive pulmonary blood flow preoperatively may have postoperative pulmonary hypertension requiring pulmonary vasodilators, aggressive sedation, and paralytics
- Extubation timing depends on complexity of repair and whether delayed sternal closure is required

Lesion Specific Postoperative Concerns

- DORV with subaortic or doubly committed VSD with PS
 - Residual VSD or LV outflow tract obstruction
 - Tricuspid regurgitation
 - LV dysfunction
- DORV with subpulmonary VSD (Taussig-Bing)
 - Coronary injury
 - Neoaortic/neopulmonary obstructions
 - Residual VSD
 - REV: pulmonary insufficiency
- DORV with subaortic VSD without PS
 - Subaortic obstruction
 - Pulmonary outflow tract obstruction
 - Residual VSD
 - CHB is rare

- **DORV with doubly committed VSD**
 - Tunnel dehiscence or obstruction
 - Residual VSD
 - RVOT obstruction
 - Subaortic stenosis
 - Complete heart block
- **DORV with non-committed VSD**
 - Aortic outflow tract obstruction
 - Pulmonary outflow tract obstruction
 - Residual VSD

CHAPTER 12

Heart Failure

Chapter Editors

Mary McBride, MD

Lindsay Jackson, APN, CPNP-AC

Chapter 1: Heart Failure

Chapter 2: Cardiomyopathy

Chapter 3: Myocarditis

Section 1 Heart Failure

Adam Lowry, MD and Sherrill Gutierrez, MD

Etiology

- Primary cardiomyopathy
 - Dilated
 - Hypertrophic
 - Restrictive
 - Non-compaction
 - Arrhythmogenic right ventricular cardiomyopathy (ARVC)
- Secondary cardiomyopathy

Table1. Selected Etiologies

Cardiac	<u>Ischemic</u> : coronary (i.e., ALCAPA), coronary calcinosis, Kawasaki, familial hyperlipidemia <u>Valvular</u> : LVOTO, MR (multiple etiologies) <u>Arrhythmia</u> : active or remote, atrial or ventricular HTN
Infectious	Viral (including myocarditis) Chagas Disease, rheumatic fever
Autoimmune	Systemic lupus erythematosus, polyarteritis nodosa, rheumatoid arthritis, scleroderma, juvenile dermatomyositis
Metabolic	<u>Endocrine</u> : thyroid, adrenal Insufficiency, pheochromocytoma, acromegaly, diabetes mellitus, <u>Infiltrative</u> : iron, Wilson's, amyloidosis <u>Dietary</u> : carnitine (1° or 2°), selenium
Inborn Errors of Metabolism	<u>Amino Acid</u> : homocystinuria, alkaptonuria, oxalosis, hyperglycinemia <u>Mitochondrial</u> : resp chain, cytochrome-c enzymes, MELAS, MERRD, mitochondrial myopathies <u>Storage</u> : glycogen storage diseases, mucopolysaccharidoses, mucopolipidoses, gangliosidoses, lipid storage, Gaucher's, Fabry's
Neuromuscular	Duchenne/Becker dystrophies, Friedreich ataxia
Toxin	Anthracyclines (chemo), radiation, alcohol
Peripartum	

Pathophysiology

- Syndrome resulting from a combination of neuro-hormonal dysregulation and systolic/diastolic myocardial pump failure

- Decreased cardiac output leads to up regulation of neuro-hormonal pathways
 - Increased renin-angiotensin-aldosterone system leads to increase sodium and water retention
 - Increase preload and myocardial stretch
 - Pulmonary venous congestion and pulmonary edema
 - Activation of the sympathetic nervous system
 - Tachycardia increases metabolic demand of myocardium with decreased function
 - Increased systemic vascular resistance causing increase afterload to impaired myocardium

Physical Exam

- High index of suspicion needed to make accurate diagnosis
- Comprehensive physical exam must include assessment for the following
 - Cardiac: tachycardia, gallop, arrhythmia, murmurs
 - Pulmonary: crackles, diminished breath sounds, tachypnea
 - Perfusion: capillary refill, pulses, skin color, skin temperature
 - Liver: hepatomegaly
 - Abdomen: ascites
- Symptoms differ based on age of the patient and underlying etiology
 - Neonates
 - Irritability, restlessness
 - Tachypnea
 - Tachycardia
 - Hepatomegaly
 - Children and adolescents
 - Vomiting
 - Tachypnea
 - Tachycardia
 - Hepatomegaly
 - Severe abdominal pain
 - Peripheral edema
- Hypotension, bradycardia, altered mental status, and respiratory failure are late signs of cardiogenic shock
- Patients with chronic compensated heart failure often present with decompensated state following intercurrent illness or as a result of natural disease progression
 - Physical exam might not reflect degree of alteration in cardiac output

Diagnostic Studies

- Imaging
 - Echocardiography
 - Rule out structural defects
 - Assess ventricular size and wall thickness, systolic function, and atrial size
 - Assess coronary anatomy and wall motion abnormalities

- MRI
 - Superior assessment of RV size and function compared to echocardiography
 - Visualization of proximal coronary artery anatomy is superior to echocardiography
 - Gold standard for diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC)
 - Use of gadolinium may help define myocardial scar (old injury or subacute injury) and areas of edema associated with acute myocarditis
- Catheterization
 - Biopsy is gold standard for diagnosis of myocarditis
 - Risk of perforation in patients with acute fulminant myocarditis and thin ventricular wall
 - Hemodynamic assessment is gold standard for diagnosis of restrictive cardiomyopathy
- ECG
 - Often abnormal, though not specific (sinus tachycardia, nonspecific T wave abnormalities, nonspecific interventricular conduction delay, partial bundle branch block)
 - Occasionally reveals underlying etiology of acute decompensated heart failure
 - Define the type of tachycardia causing cardiomyopathy
 - Inferolateral Q waves in ALCAPA
 - Severely increased precordial voltage pattern in Pompe's disease
- Lab evaluation
 - End-organ assessment
 - Liver and renal function tests, acid-base status, and CBC
 - N-terminal-pro-BNP/BNP
 - Marker of volume status and preload
 - Correlate with heart failure severity
 - Most useful when used to trend over time
 - Must consider risk/benefit of obtaining invasive testing, as well as cost/benefit of specialized testing; a sequential approach is recommended in the non-critical newly diagnosed patient

Management

- Management directed at improving congestion while optimizing oxygen delivery and reducing oxygen consumption.
- Optimize oxygen delivery
 - Diuretics
 - Decreases pulmonary congestion resulting from elevated left atrial pressure and systemic venous congestion by decreasing right atrial pressure

- Inotropic support
 - Use of inotropic agents should be based on clinical assessment and not echocardiogram or other ancillary test/studies
 - Optimize chronotropy and inotropy to increase cardiac output
 - Increases risk of arrhythmias and increases myocardial oxygen consumption
 - No studies have demonstrated improved survival with use of inotropic agents
- Vasodilators
 - Decreases systemic vascular to increase cardiac output and decrease congestion
- Mechanical ventilation
 - Decreases transmural pressure which is a highly effective means of reducing left ventricular afterload
 - Positive pressure ventilation increases right ventricular afterload which may impair cardiac output
- Decrease metabolic demand
 - Sedation/neuromuscular blockade
 - Mechanical ventilation
 - Decreases metabolic demand and oxygen consumption
 - High risk procedure which can precipitate cardiac arrest or malignant arrhythmias
 - Avoid fever/tachycardia
- Anticoagulation
 - Reserved for patients with severely decreased function and/or evidence of thrombus on echocardiography to minimize stroke risk
- Advanced therapies for refractory decompensated heart failure
 - Mechanical circulatory support may be considered for decompensated heart failure refractory to medical management
 - Indication, patient size, clinical urgency, anticipated duration of support, and heart transplant candidacy contribute to decision making

Section 2
Cardiomyopathy
Tyler Harris, MD

Dilated cardiomyopathy (DCM)

Pathophysiology

- Most common cause of congestive heart failure in children without known structural abnormalities
- Typically idiopathic without identifiable cause at presentation
- Primary usually a result of genetic abnormality in cardiac myocytes
- Secondary due to myocardial insult from variable source
- Ventricular dilation, impaired systolic function, and contractility
- Ventricular wall may be normal or thinned

Presentation

- Signs and symptoms consistent with congestive heart failure
- If due to metabolic/toxic cause symptoms will occur at times of increased demand (fever, illness, fasting) and patients often present with concomitant signs of hypoglycemia, metabolic acidosis, and encephalopathy

Diagnostic Evaluation

- ECG
 - Nonspecific changes including increased left ventricular forces, flat/inverted T waves, atrial enlargement
 - Tachyarrhythmia, sinus tachycardia
- Echocardiography
 - Quantify systolic and diastolic function, LV end-diastolic dimension, and LV volume
 - Patients typically present with global systolic dysfunction, but may have segmental wall motion abnormalities
 - Evaluate for AV valve regurgitation
 - Evaluate for underlying anatomic causes including outflow obstruction, coarctation, primary valvular disease, and coronary anomalies
 - Look for thrombi in patients with ejection fraction ~20% or less
- Magnetic resonance imaging
 - Most useful if considering the diagnosis of acute myocarditis
 - Focal wall motion abnormalities and delayed enhancement
- Cardiac catheterization
 - May be useful to assess coronary arteries

Management

- Decrease congestion and optimize oxygen delivery/cardiac output
 - Diuretic therapy
 - Inotropy

- Should be used based only on clinical assessment of cardiac output to improve contractility
- Cause increase myocardial oxygen consumption
- Proarrhythmic
- Antiarrhythmic agents
 - Only when arrhythmia is present
 - Caution because of propensity to depress contractility
- Afterload reduction
 - Vasodilator therapy
 - Positive pressure ventilation
- Optimize oxygen carrying capacity
 - Oxygen
 - Blood transfusion when indicated
- Decrease oxygen consumption
 - Sedation and neuromuscular blockade
 - Parenteral nutrition to limit splanchnic blood flow
 - Avoid fever, seizures
- Anticoagulation in presence of severely decreased function or known intramural thrombus
- Mechanical circulatory support for patients unresponsive to medical therapy

Hypertrophic Cardiomyopathy

Pathophysiology

- Variable presentation/natural history
- Associated with abnormal myocyte orientation and fibrosis
- Symptoms result from diastolic dysfunction and outflow tract obstruction

Presentation

- Signs of diastolic dysfunction usually with preserved or hyperdynamic systolic function
 - Dyspnea on exertion
- Syncope with exertion, angina or ventricular arrhythmias as a result of LV outflow obstruction and/or progressive hypertrophy
- Physical exam
 - Laterally displaced and forceful LV impulse
 - Loud S3/S4
 - Systolic ejection murmur at right upper sternal border

Diagnostic Evaluation

- Family history
 - Sudden cardiac death in the family
- ECG
 - Left ventricular hypertrophy with or without strain pattern
 - Abnormal QRS axis with presence of Q waves

- Echocardiogram
 - Asymmetric hypertrophy (septum>free wall)
 - Preserved systolic function and volume
 - Decreased diastolic function
 - Outflow tract obstruction due to septal hypertrophy and systolic anterior motion of the mitral valve in ~20% of patients

Management

- Decrease outflow obstruction
 - Avoid tachycardia
 - Beta blockers
 - Adequate preload
 - Treat secondary causes of tachycardia
 - Myomectomy or sympathectomy used for refractory cases
- Management of diastolic dysfunction
 - Diastolic filling time
 - Treat arrhythmias or sinus tachycardia
 - Antihypertensive therapy may be beneficial
- Indications for implantable cardiac defibrillator
 - Syncope
 - Ventricular arrhythmias
 - Massive hypertrophy
 - Sudden cardiac death in first degree relative

Restrictive Cardiomyopathy

Pathophysiology

- Symptoms result from diastolic dysfunction, restricted ventricular filling, and atrial enlargement
- Normal systolic function, ventricular volume, and thickness
- High filling pressures with low CO
- Primary restrictive cardiomyopathy is a result of a gene defect
- Secondary causes include infiltration/injury as seen in connective tissue disorders, medication induced injury (anthracyclines), radiation, neoplasms, and hemochromatosis
- Progressive disease with poor prognosis

Presentation

- Variable and age dependent
- Failure to thrive common presenting sign in neonates and infants with recurrent respiratory infections from pulmonary congestion
- Chronic congestion
 - Pulmonary edema, ascites, edema, and hepatomegaly
- Syncope (high mortality)
- Arrhythmias

- Physical exam
 - Poor growth
 - Signs of elevated biventricular diastolic pressure
 - Pulmonary edema and congestion with cough and tachypnea
 - Jugular venous distension
 - Ascites, hepatomegaly

Diagnostic Evaluation

- Chest x-ray
 - Cardiomegaly
 - Diffuse pulmonary congestion
- ECG
 - Biatrial enlargement
 - ST/T wave changes
 - Atrial arrhythmias
- Echocardiography
 - Normal ventricular dimensions with biatrial enlargement (Mickey Mouse ears)
 - Abnormal diastolic filling and inflow patterns on doppler evaluation
 - Elevated pulmonary artery pressure secondary to left atrial hypertension
 - Dilated hepatic veins
- Cardiac catheterization
 - Significant elevated end diastolic pressure in left and right ventricles
 - Significant elevation end diastolic pressures with fluid challenge

Management

- Goal is to optimize growth and hemodynamics while evaluating candidacy for transplantation
- Decrease congestion
 - Diuretics
 - Must be used with caution because of preload dependence
- Optimize diastolic filling
 - Avoid tachycardia, may require antiarrhythmic therapy
- Transplantation
 - Early transplantation should be considered in all patients given prognosis and progressive nature of the disease

Section 3
Myocarditis
Mary E. McBride, MD

Pathophysiology

- Inflammation of the myocardium with necrosis which is associated with impaired ventricular function
- Etiologies include systemic diseases, drugs, toxins, bacterial, fungal, and viral infections
 - Viral causes are most common

Presentation

- May present with signs of congestive heart failure
 - Tachycardia, hypotension, metabolic acidosis, syncope, arrhythmia, chest pain, peripheral edema, jugular venous distension, hepatomegaly, and shortness of breath
- Non-specific upper respiratory or gastrointestinal complaints
- Fulminant versus indolent
 - Fulminant myocarditis typically present with acute decompensated heart failure and often require inotropic support and/or mechanical circulatory support
 - Indolent cases typically present with signs of mild congestive heart failure and congestion

Diagnostic evaluation

- Biomarkers
 - Troponin
 - Lactate
 - BNP
- ECG
 - Nonspecific changes
 - Low voltage, ST, T wave changes, tachycardia, bundle branch block, heart block, and tachyarrhythmias
- Echocardiography
 - Left ventricular dilatation and dysfunction
 - Global dyskinesia
 - AV insufficiency
- Cardiac catheterization
 - Gold standard for diagnosis
 - High rate of sampling error given patchy nature of pathology

- Specimens analyzed to look for fibrosis, inflammation, T-cells, viral capsids, viral antigens, and PCR
- Balance with risks of procedure and anesthesia with diagnostic benefit
- **Magnetic resonance imaging**
 - Presence of late enhancement after gadolinium contrast injection is helpful in diagnosis of myocarditis
 - Balance with risks of anesthetic needs with diagnostic benefits

Management

- Supportive care remains the standard with the goal of optimizing oxygen delivery and minimize metabolic demand
- Improve oxygen delivery
 - Diuretics should be used to manage congestion
 - Vasoactive support with beta adrenergic agents should be used with caution because of arrhythmogenicity and increase in myocardial oxygen consumption
 - Often unavoidable in patients with cardiogenic shock and acute fulminant presentation
 - Blood transfusion to optimize oxygen carrying capacity
 - Mechanical ventilation to decrease LV afterload
 - Must be done with caution because of risk for arrhythmia and sudden arrest in patients inadequately anesthetized or with sudden change in systemic vascular resistance
- Decrease metabolic demand
 - Sedation/neuromuscular blockade
 - Mechanical ventilation
 - Avoid fever/tachycardia
- Mechanical circulatory support should be considered for all patients refractory to medical management
- Intravenous immunoglobulins (IVIG)
 - There is no pediatric evidence of improved outcomes with the use of IVIG
- Anticoagulation
 - Consider anticoagulation for patients with severely decreased ejection fraction or evidence of thrombus on echocardiogram

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CHAPTER 13

Heart Transplantation

Chapter Editors

Matthew O'Connor, MD

Carley Bober, MSN, CRNP

Section 1: Listing and Preoperative Considerations

Section 2: Operative and Postoperative Management

Section 3: Immunosuppression

Section 4: Rejection

Section 1
Listing and Preoperative Considerations
Sherry Pye, MNSc, APRN, CCRN, CCTC

Transplant Evaluation Process

- Indications for transplantation: heart transplantation remains the definitive therapy for end-stage heart failure in children
- Generally accepted indications for transplant include
 - Physical limitations due to systemic ventricular dysfunction not amenable to medical or surgical therapy
 - Growth failure associate with systemic ventricular dysfunction
 - Restrictive cardiomyopathy with reactive pulmonary vascular resistance
 - Specific anatomic defects (with or without cyanosis) that are not amenable to further surgical or medical interventions
- There are few, if any, absolute contraindications to HT
- Relative contraindications in children include
 - Severe irreversible multi-organ disease.
 - Fixed elevations in pulmonary vascular resistance, generally > 8 Wood units indexed to body surface area (BSA)
 - Severe hypoplasia of the central pulmonary arteries and/or pulmonary veins
 - History of another medical condition which limits life expectancy greater than expected graft survival (i.e., active malignancy, recent HIV or hepatitis infection)
 - History of tobacco and/or illicit substance abuse
 - Severe psychological or behavioral problems in the patient or patient's caregivers that may limit adherence to post-transplantation care regimens
- Components of the evaluation: the transplant evaluation is a detailed, multidisciplinary process by which a potential candidate's suitability for transplant is assessed. Consent/assent from the patient and/or patient's guardian are required prior to initiation of evaluation. Basic components include
 - Formal family conference outlining the current medical condition and specific details of transplant including the evaluation process, surgical procedure, and any anticipated complications or issues at the time of transplant, expected postoperative care and recovery period, risks of transplant and immunosuppression, medical or surgical alternatives to transplant, and absolute requirement for patient/family adherence and commitment to life style changes after transplant
 - General physical examination, measurement of height, weight, and vital signs
 - Medical and surgical history including specific surgical cardiac procedures

- Two blood typing samples drawn at a different times
- HLA typing for future donor specific crossmatch
- Testing for specific viral diseases: CMV, EBV, HCV, HIV
- Evaluation of all systemic metabolic parameters such as renal function, liver function, and thyroid function.
- In selected situations, CT scan or MRI of the brain to assess for any neurologic abnormality
- In selected cases, complete abdominal ultrasound to assess status of liver, gallbladder, and kidneys
- In selected situations, cardiac catheterization to assess current hemodynamic parameters and pulmonary vascular resistance
- Complete dental evaluation to determine current dental health and need for any restoration therapy prior to transplant
- Review of immunization record
- Psychosocial evaluation of patient and family support systems, financial resources, history of tobacco or drug use and any other psychosocial issue that may limit adherence to medical regimen
- Financial guarantee with approval obtained by insurance prior to listing and transplant procedure

Listing Process

- Once the evaluation process is complete, the multidisciplinary team must make the clinical decision to proceed with (or decline a candidate for) transplant listing
- Pediatric patients in the United States are listed for transplant through the United Network for Organ Sharing (UNOS) by blood type, weight, and clinical severity criteria
- Organ allocation
 - Severity status (organs are offered first to 1A candidates, then 1B candidates, then status 2 candidates)
 - Blood type
 - Accumulated wait time
 - Geographic location
 - Age (pediatric donors are offered to pediatric candidates first)
- UNOS waitlist categories
 - Status 1A
 - Requires high dose single inotrope therapy or multiple inotropes, mechanical ventilation or mechanical circulatory support
 - Status renewed every 2 weeks
 - Status 1B
 - Requires low dose inotropes or demonstrates growth failure
 - Status 2
 - Does not meet status 1A or 1B criteria
 - Status 7

- Inactive status: deemed temporarily not a candidate for transplantation (i.e., intercurrent systemic infection) until the condition causing the unsuitability for transplantation resolves
- Patients with severe disease not meeting specific 1A or 1B criteria may receive an exception for 1A or 1B status pending case review by a multicenter review board
- Waitlist duration: pediatric wait list duration is affected by many factors
 - Blood type (O blood type patients wait longer)
 - Age
 - Weight
 - Geographic region
 - HLA sensitization
 - Underlying diagnosis (cardiomyopathy vs congenital heart disease)
- Waitlist outcomes
 - Donor availability is the key factor that dictates waitlist time
 - Waitlist mortality is nearly 20%
 - Risk factors for waitlist mortality include: 1A status, ECMO support, dialysis, mechanical ventilation, congenital heart disease, non-white ethnicity, and renal insufficiency

Special Situations

- ABO incompatible transplantation
 - Possible in young children because antibodies to non-self ABO antigens (isohemagglutinins) generally do not develop until 1-2 years of age
 - Candidates <2 years of age have serum anti-A and anti-B antibody-titers (isohemagglutinins) drawn at time of evaluation
 - Candidates not be eligible to receive an ABO incompatible transplant if the anti-A or anti-B titers are unacceptably high
 - Triple volume exchange transfusion is performed at time of transplant
 - Outcomes are similar to those who receive compatible grafts, although long-term (>10 years) follow up data are lacking
- The sensitized patient
 - Assessment for development of anti-HLA antibodies occurs at each evaluation
 - HLA antigens are found on nearly all cells in the body and make up an individual's immune system fingerprint
 - Anti-HLA antibodies are antibodies against non-self HLA antigens and may develop from blood transfusions, pregnancy, receipt of allograft tissue at surgery, and mechanical circulatory support
 - Quantification of anti-HLA antibody burden is done via panel reactive antibody (PRA)

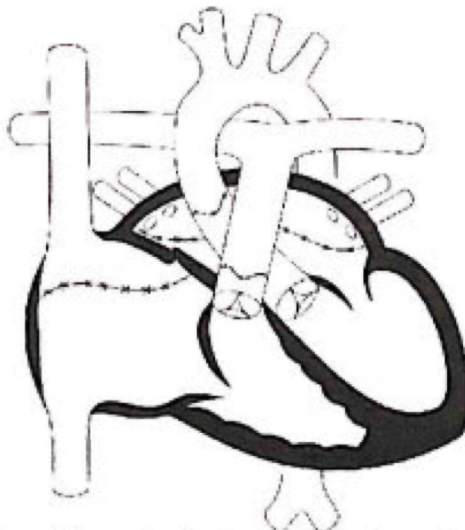
- In general >10% for class I and/or class II anti-HLA antibodies defines clinically significant production in a recipient and places the patient at higher risk of rejection
- HT can proceed despite the presence of HLA antibodies in the recipient, but may require longer waitlist time and/or enhanced immunosuppression at the time of transplantation
- Desensitization therapies such as plasmapheresis and IVIG have been used during the perioperative period
- In utero listing
 - Candidate may be listed for transplant while in utero however it is uncommon
 - Generally reserved for conditions in which there is an expectation of very high cardiac-related mortality during the first month of life
- Heart-lung transplantation
 - Reserved for patients with end-stage heart disease and fixed, non-reactive elevations of pulmonary vascular resistance.
 - Potential heart-lung recipients undergo a similar evaluation process to those undergoing heart transplant
 - Outcomes of heart-lung transplantation are much less favorable than heart transplant alone

Preoperative Considerations

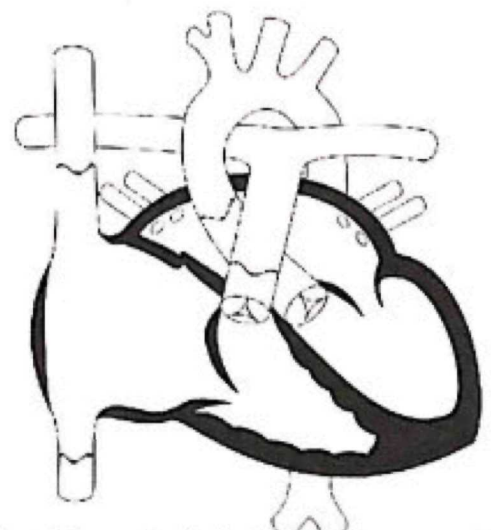
- Donor history
 - Duration of cardiopulmonary resuscitation
 - History of chest wall trauma (cardiac contusion)
 - CDC defined risk factors that increase risk of transmission of infectious agents to the recipient (i.e., sexual history, IV drug use, transfusion history)
 - Donor size in relation to recipient: in general, donors 80% to 200% of the recipient weight can be successfully transplanted
- Recipient factors
 - Patients with complex congenital anatomy require additional dissection time in the operating room prior to transplant and may need complex and/or creative systemic venous anastomoses and arch reconstruction
 - Patients supported with VAD prior to transplant may require additional time in OR prior to transplant for dissection and device removal
 - Use of anticoagulation may increase risk of intraoperative and postoperative bleeding

Operative Procedure

- Operative technique
 - Surgical technique is either biatrial or bicaval anastomosis
 - In a biatrial anastomosis, "cuffs" of the recipient left and right atrium are left in situ
 - In a bicaval anastomosis, only a left atrial cuff is present and the venous anastomoses are located on the IVC and SVC



Heart Transplant - Bi-atrial Anastomosis



Heart Transplant with Bicaval Anastomosis

- Factors impacting function of new heart
 - Ischemic time: time elapsed between aortic cross clamping of the donor heart at procurement and release of the aortic cross clamp in the recipient after implantation of the heart
 - Ischemic times of greater than 4 hours are associated with decreased ventricular function and worse outcomes
 - Administration of induction therapy/immunosuppression
 - Agents used are center-specific
 - Generally, high dose corticosteroids and antimetabolites (mycophenolate mofetil) are given prior to release of the aortic cross clamp.
 - Cardiopulmonary bypass time
 - Intraoperative complications: bleeding secondary to extensive suture lines, right ventricular dysfunction secondary to elevated PVR, ventricular dysfunction due to prolonged ischemic time or size discrepancy, arrhythmias, and hyper-acute rejection

Postoperative Considerations and Management

- Right ventricular dysfunction due to elevated PVR is common in patients with a preoperative diagnosis of dilated cardiomyopathy, right sided heart failure or mitral stenosis
- iNO may be used to reduce RV afterload
- Temporary pacing and/or selective agonist infusions (isoproterenol) may be used in the early postoperative period to optimize heart rate and augment cardiac output
- Inotropic agents may be used in the early postoperative period to support ventricular function
- May require magnesium and calcium supplementation
- Early postoperative immunosuppression regimens vary among centers depending on induction regimen, presence of anti-HLA sensitization, renal function, medication levels, CD3 counts, and other recipient-specific factors
- Conversion to oral agents should occur quickly to avoid sequelae of IV agents and achieve therapeutic levels early in the postoperative course
- HLA crossmatch should be available within 12-24 hours from transplantation
 - If positive crossmatch (recipient has donor-specific antibodies), immunosuppression may be augmented and/or the patient may undergo antibody-depletion therapies such as plasmapheresis
- Long-term IV access may be required if serial antibody-depletion therapy required
- Surveillance for infection and rejection is critical
- Pan-cultures should be sent with any fever and antibiotic coverage may be initiated or broadened

- Oral antifungal agent and PCP prophylaxis should be started as soon as patient able to tolerate oral or enteral feeds
- Ganciclovir is administered to patients at risk for CMV (donor or recipient positive)
- Serial ECGs may be obtained to screen for signs of rejection
- Patients are routinely screened for CMV and EBV using PCR

Transplant Specific Postoperative Concerns

- Sinus node dysfunction and bradyarrhythmias
- Bleeding especially if extensive dissection required or preoperative history of anticoagulation
- May be at risk for transfusion related acute lung injury due to exposure to multiple blood products
- Low cardiac output syndrome due to ventricular dysfunction in patients with longer ischemic times, marginal donor heart or narrowing at anastomosis site
- The pulmonary arteries may not be well seen by echocardiography, a low threshold for cardiac catheterization should be maintained
- Right ventricular dysfunction in patients with history of elevated PVR or prolonged CHF
- Donor right ventricle is may not tolerate high afterload due to elevated pulmonary vascular resistance
- Systemic inflammatory response syndrome especially in patients requiring prolonged cardiopulmonary bypass times
- Rejection: (see Section 4)
- High risk for acute kidney injury in the setting of low cardiac output and nephrotoxic medications

Section 3
Immunosuppression
Kelci Laporte, RN, CPNP-AC, CCTC

Types of Immunosuppression

- Induction is given at the time of transplant
- Maintenance: typically a 2-drug regimen of calcineurin inhibitor and antimetabolite
- Augmentation of treatment for rejection: typically includes pulse steroids and/or T-cell depletion

Classes of Immunosuppressive Medications

- T-cell specific therapies (ATG, ATGAM, OKT3)
 - Typically used for induction, can also be used for treatment of rejection
 - Mechanism of action: disruption of the T-cell receptor leading to loss of function of T cells and removal from peripheral blood
 - Side effects: anaphylaxis leukopenia, thrombocytopenia, arthralgia
 - Pros: highly effective, reversal of rejection
 - Cons: rare side effect of cytokine release syndrome (most frequent with OKT3), can increase risk of CMV infection and risk of post-transplant lymphoproliferative disorder (PTLD)
- IL-2 receptor antagonists
 - Typically used for induction
 - Mechanism of action: monoclonal antibody which blocks IL-2 receptor complex (required for activation of T-cells and B-cells)
 - Side effects are uncommon, may include hypersensitivity
 - Pros: low side effect profile
 - Cons: does not deplete T-cells so not as durable an immunosuppressant as T-cell antibodies
- Corticosteroids (prednisone, methylprednisolone)
 - Standard first line rejection treatment
 - Mechanism of action: involved with regulation of genes, inflammation, and lymphocyte effector activity
 - Directly toxic to lymphocytes
 - Dosing
 - Maintenance: dosing regimen is center specific.
 - Many centers wean patients off steroids shortly after transplant, long term use of low dose steroids may be reasonable for patients with history of recurrent or humoral rejection
 - Rejection: typically 10-15 mg/kg/day IV x3 days
 - Side effects: hypertension, diabetes mellitus, growth retardation, and hyperlipidemia
 - Pros: can be used to treat rejection as an outpatient

- Cons: long-term use can cause chronic adrenal insufficiency and increased incidence of opportunistic infections
- Calcineurin inhibitors (tacrolimus (FK506, Prograf), cyclosporine)
 - Mechanism of action
 - Tacrolimus: blocks calcineurin which is necessary for production of cytokines (particularly IL-2) that are needed for production and proliferation of cytotoxic T-cells
 - Cyclosporine (CSA): similar to tacrolimus however differs by binding to a different protein which inhibits calcineurin
 - Typical maintenance dosing
 - Tacrolimus: 0.1 to 0.3 mg/kg/day divided every 12 hours
 - Cyclosporine: 2mg/kg/dose every 12 hours
 - Side effects: nephrotoxicity, hypertension, hyperlipidemia, diabetes mellitus (tacrolimus), gingival hyperplasia (CSA)
 - Pros: effective, used as first line for maintenance immunosuppression
 - Cons: chronic renal insufficiency, increased incidence of PTLD
 - Therapeutic monitoring of serum blood levels
 - Trough tacrolimus level 6-10ng/mL
 - Trough CSA level 200-400ng/mL
- Antimetabolites (mycophenolate mofetil (CellCept), azathioprine (Imuran))
 - Mechanism of action: block DNA and RNA synthesis in lymphocytes by inhibiting de novo purine synthesis
 - Typical maintenance dosing
 - Mycophenolate mofetil: 600mg/m²/dose every 12 hours
 - Azathioprine: 2-4 mg/kg/day once daily
 - Side effects: GI distress (mycophenolate), myelosuppression (more common with azathioprine)
 - Pros: when combined with tacrolimus it is very effective at preventing rejection
 - Cons: use may be limited due to side effects, mycophenolate is teratogenic
 - Therapeutic monitoring: serum levels of mycophenolate can be obtained but are of limited clinical use
 - Monitor myelosuppression with CBC and differential
- mTOR inhibitors (sirolimus (Rapamycin))
 - Mechanism of action: sirolimus inhibits target of mammalian rapamycin (mTOR) which prevents T-cell and B-cell proliferation and differentiation in response to cytokines such as IL-2, inhibits proliferation of smooth muscle in the vasculature
 - Maintenance dosing: 1mg/m² once daily, often given in combination with attenuated dose of tacrolimus
 - Side effects: mouth sores, hyperlipidemia, hypertriglyceridemia, myelosuppression, and poor wound healing

- Pros: may help slow progression of coronary vasculopathy, once daily dosing is generally not nephrotoxic
- Cons: inhibits wound healing, possible risk of rejection as monotherapy
- Therapeutic monitoring: serum blood levels measured by 24 hour trough, monitor liver function and CBC during treatment

Section 4
Rejection
Sherry Pye, MNsc, APRN, CCRN, CCTC

Rejection

- Immunologic response process by which non-self endothelial cells on the transplanted heart are recognized and targeted for destruction by the recipient's circulating lymphocytes and/or antibodies
- Complex process which may be cellular (T-cells) or antibody (B-cells) mediated can occur at any time following transplant
- Rejection is classified into four categories

Hyperacute Rejection (Rare)

- Catastrophic immune response that typically occurs immediately (minutes to hours) following transplant
- Usually seen only in the setting of inadvertent ABO incompatibility or presence of high-titer, pre-formed anti-HLA antibodies
- Frequently leads to graft loss and urgent listing for retransplantation

Acute Cellular Rejection (ACR)

- T-cell mediated immunologic response that may occur at any time following transplant
- Highest risk of ACR is within the first 6 months following transplant
- Approximately 20% of patients will experience ACR within the first year
- Beyond the first year, episodes of ACR are often associated with non-adherence to the immunosuppression regimen
- Diagnosed by endomyocardial biopsy and graded according to a standard scale proposed by the International Society of Heart & Lung Transplantation (ISHLT)
- 2004 ISHLT Grading Scale
 - 0R: indicates no evidence of ACR
 - 1R: mild—indicates interstitial and/or perivascular infiltrate with up to 1 focus of myocardial damage
 - 2R: moderate—indicates 2 or more foci of infiltrate with associated myocyte damage
 - 3R: severe—indicates diffuse infiltrate with multifocal myocyte damage with or without edema, hemorrhage, or vasculitis

Antibody-Mediated Rejection (AMR)

- Graft damage by donor specific antibodies directed against donor graft HLA antigens
- Incidence may be higher than previously appreciated due to a larger number of undiagnosed, asymptomatic patients
- Risk factors include high titer pre-transplant HLA antibodies, incompatible HLA crossmatch, and development of "de novo" donor specific antibodies

- Diagnosed by formal histologic and immunopathologic criteria proposed by the ISHLT on endomyocardial biopsy specimen
- ISHLT Grading Scale
 - pAMR 0: indicates histologic and immunopathologic studies are both negative
 - pAMR 1 (H+): indicates histologic findings are present and immunopathologic studies are negative
 - pAMR 1 (I+): indicates histologic findings are negative and immunopathologic studies are positive (CD68+ and/or C4d+)
 - pAMR 2: indicates both histologic findings and immunopathologic studies are positive
 - pAMR 3: indicates interstitial hemorrhage, capillary fragmentation, mixed inflammatory infiltrates, endothelial cell pyknosis and/or karyorrhexis, marked edema, and immunopathologic findings are present

Chronic Rejection

- Recurrent, refractory rejection despite aggressive medical monitoring and management along with adherence to immunosuppression regimen
- Risk factors: African-American or Hispanic race, prior incidences of rejection and shorter period between episodes of rejection
- Leads to early graft dysfunction and loss due to progressive coronary allograft vasculopathy (CAV)
- CAV is defined as chronic intimal growth of the coronary artery lumen leading to diffuse, progressive narrowing of the artery and decrease in myocardial perfusion

General Signs and Symptoms of Rejection

- Often vague and nonspecific
- Pericardial effusion/friction rub, decreased QRS voltages on ECG, cardiomegaly on CXR, arrhythmias or conduction disturbances, fatigue, low grade fever, gallop rhythm, pulmonary changes of crackles or wheezing, weight gain, decreased graft function (systolic or diastolic) on echocardiogram, hemodynamic instability with hypotension and end-organ dysfunction
- Gold standard for diagnosis is endomyocardial biopsy
- Successful treatment of rejection is dependent on early recognition and treatment to prevent the graft damage and/or death

Treatment of Rejection

- Presenting signs and symptoms and ISHLT Grading Scale will dictate scope of therapy required including whether treated as inpatient versus outpatient for Grade 1R
- Acute cellular (ACR)
 - May be initiated empirically before the patient undergoes biopsy
 - Corticosteroids are the mainstay of treatment for ACR

- IV corticosteroids and hospitalization instituted for Grade 2R or higher
- May require inotropic support and close monitoring for hemodynamic changes or symptoms that indicate low cardiac output
- Follow up cardiac catheterization and biopsy should occur 2-3 weeks following rejection (depending on age/size of child) to monitor for graft improvement and histologic resolution of ACR
- May require increase in baseline immunosuppression
- Refractory ACR may require T-cell depleting agents such as antithymocyte globulin (ATG)
- Antibody-mediated (AMR)
 - Preventative measures are key due to poor outcomes associated with AMR
 - High levels of preoperative anti-HLA antibodies predict the risk for development of postoperative AMR
 - Difficult to manage and often includes plasmapheresis, photopheresis, intravenous immune globulin or targeted anti-B cell therapies such as rituximab or bortezomib along with augmentation of standard immune suppression
 - Avoidance HLA antigens
- Chronic rejection
 - Proactive avoidance of risk factors
 - Consider additional immunosuppressive medications such mTOR inhibitors with specific effects against CAV
 - HMG-Co-A reductase inhibitors (pravastatin) may slow progression of CAV
 - Retransplantation is the only effective long-term therapy for patients who have developed CAV and graft dysfunction

Proposed Table 1 – Commonly Used Immunosuppressive Medications

Drug Class	Specific Medications	Usual Dose	Therapeutic Monitoring	Adverse Effects
Corticosteroids	Prednisone (PO) Methylprednisolone (IV)	Varies depending on use; usually 1-2 mg/kg/day PO for maintenance therapy; 10-20 mg/kg/day for induction/rejection	None	Poor wound healing, hypertension, hyperglycemia, adrenal suppression, bone demineralization
Calcineurin Inhibitors	Cyclosporine Tacrolimus	Cyclosporine: 4 mg/kg/day PO divided BID-TID Tacrolimus: 0.1 – 0.3 mg/kg/day PO divided BID IV dosing available but uncommon	Trough (30 min before 12 hour dose) levels: Cyclosporine 200-400 ng/mL Tacrolimus 6-10 ng/mL	Cyclosporine: diabetes mellitus, gingival hyperplasia Tacrolimus: nephrotoxicity Both: many drug interactions
mTOR Inhibitors	Sirolimus	1 mg/m ² daily PO	Trough (30 min before 24h dose: 5-15 ng/mL)	Poor wound healing
Antimetabolites	Mycophenolate mofetil Azathioprine	Mycophenolate mofetil: 600 mg/m ² /dose BID IV/PO Azathioprine: 2-4 mg/kg/day PO	CBC with differential	Mycophenolate mofetil: GI distress Azathioprine: cytopenia
T-Cell Specific Medications	ATG, ATGAM	Varies	CD3 counts	Cytopenia, possible increase in risk for lymphoma
B-Cell Specific Medications	Rituximab, Bortezomib	Varies	CD19, CD20 counts	Cytopenia, possible increase in risk for lymphoma
Other	Basiliximab (IL-2 receptor antagonist)	Varies	None	Hypersensitivity reactions

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CHAPTER 14

Mechanical Circulatory Support

Chapter Editors

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Section 1: Extracorporeal Membrane
Oxygenation (ECMO)

Section 2: Ventricular Assist Devices (VAD)

Section 1

Extracorporeal Membrane Oxygenation (ECMO)

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Introduction

Extracorporeal membrane oxygenation (ECMO) provides cardiopulmonary support in conditions causing severe cardiac or respiratory failure refractory to conventional therapies

ECMO Circuit

- Venous drainage and arterial return cannulae
- Bladder (optional)
- ECMO pump (centrifugal or roller)
- Oxygenator (hollow fiber or membrane)
- Heat exchanger

ECMO Modes

- Veno-Arterial ECMO (VA ECMO): blood is drained into the ECMO circuit from the venous circulation and returned into the arterial circulation. VA ECMO can be used for cardiac and respiratory support
- Veno-Venous mode (VV ECMO): blood is drained into the ECMO circuit from the venous circulation and returned to the venous circulation. VV ECMO relies on preserved native cardiac function to maintain cardiac output. VV ECMO is thus used to support patients with isolated respiratory failure

ECMO Indications and Contraindications

- ECMO may be used when cardiac or respiratory failure become refractory to conventional therapies and the patient is at imminent risk of mortality
- ECMO can be used in all ages and is not limited by size
- ECMO indications
 - Respiratory failure (ARDS, infectious, other causes)
 - Cardiac failure (myopathies, acquired heart disease, congenital heart disease)
 - Support of failed cardiopulmonary resuscitation (i.e., extracorporeal-CPR or ECPR)
 - Procedural support for critically ill children (airway surgery, cardiac catheterization)
 - Bridge to lung or heart transplantation
- ECMO contraindications are relative and vary widely amongst ECMO programs
 - Poor prognosis from primary illness
 - Uncontrollable bleeding

- Neurological injury (severe irreversible brain injury, major intracranial hemorrhage)
- Prematurity (< 34 weeks corrected gestational age)
- Small size (< 2 kg)

Timing of ECMO Initiation

- Timing of ECMO initiation varies widely between ECMO centers and there are no established criteria for initiating ECMO
- ECMO is generally deployed when conventional therapies fail, however it should preferably be initiated prior to onset of permanent end-organ dysfunction

General Guidelines for ECMO Initiation

- After optimized conventional medical therapy
 - Continued deterioration in cardiac or respiratory function
 - Evidence of inadequate tissue oxygenation (increasing serum lactate, persistent metabolic acidosis, increasing arterio-venous oxygen content difference)
 - End-organ dysfunction (decrease in urine output, rise in serum creatinine, or liver enzymes)
 - Imminent cardiac arrest
- Criteria for ECMO initiation in neonatal respiratory failure
 - Oxygenation index > 35 for 0.3–6 hours
 - Alveolar-arterial O₂ difference > 605 mmHg for 4–12 hours
 - PaO₂ < 35–60 mmHg for 2–12 hours
 - Acidosis and shock: pH < 7.25 for 2 hours or hypotension

ECMO Team Composition

- Composition of ECMO teams may vary based on center
- ECMO systems are complex and require clear communication and interaction of all team members to provide safe and effective ECMO care
- An ideal ECMO team should include
 - Cannulating surgeon
 - ICU physician
 - Nursing staff (patient care)
 - ECMO specialist (ECMO circuit care)
 - Respiratory Therapist
 - Pharmacist
 - Blood bank personnel
 - Consulting services: Hematology, Neurology, Infectious Disease, other services
 - Physical therapy and rehabilitation services

Overview of ECMO Equipment

Types of ECMO Pumps

- Roller occlusive pump: flow set by occlusion, RPMs, and tubing size
 - Pros: can flow against high SVR or with small arterial cannulas, can be used as suction, inexpensive
 - Cons: can cause high negative and positive pressures, move air embolism to patient, and is dependent on preload to flow
- Centrifugal pump: flow set by RPMs, preload, and afterload
 - Pros: sensitive to preload and thus able to run patients at lower CVPs, unable to draw high negative pressures unless RPMs changed, air will stop pump flow
 - Cons: dependent on SVR, increased hemolysis at high negative inlet pressures, possible air entrainment at high negative pressures, retrograde flow possible

Roller pump	Centrifugal pump
Gravity drainage	Active drainage
Servo controlled	Preload and afterload controlled
Larger circuit	Smaller circuit
Bladder	No bladder
Set flow	Flow range
Complicated transports	Ease of transport

Table 1: Comparison of Roller occlusive pump and Centrifugal pump

Membrane Oxygenators

- Silicone: silicone wraps with an air bladder with blood flowing around air wrap
 - Pros: FDA approved, long term use, good air trap
 - Cons: longer de-bubbling process, no integrated heat exchanger, high transmembrane pressure
- Hollow fiber: fine silicone tubes which blood flows around
 - Pros: quick to de-bubble, integrated heat exchanger, low transmembrane pressure
 - Cons: plasma leakage noted after short term use
- Polymethylpentene (PMP): fine PMP tubes which blood flows around, true diffusion membrane
 - Pros: easy to de-bubble, integrated heat exchanger, low transmembrane pressures, no plasma leakage
 - Cons: more expensive

Cannulas

- Neck
 - Used for patients less than approximately 10 kg.
 - Easily accessible vessels during ECPR, ligation of right common carotid artery, increased chance of stroke in large patients

- Femoral
 - Used for patients greater than approximately 10 kg
 - Ease of cannulation in ECPR, percutaneous cannulation possible
 - Distal perfusion cannula if inadequate limb perfusion
 - Competing circulation between native and ECMO circulation can be noted, causing blood to the head to be more desaturated
- Transthoracic
 - Postoperative through an open sternotomy
 - Allows for very high flows as larger cannulas can be placed (i.e., patient with sepsis)
 - Increased bleeding, less secure cannulas, increased infection risk
- Confirming cannula position
 - ECMO flows
 - X-rays
 - Echocardiogram

Initial ECMO settings

- Sweep gas
 - Match membrane size to patient size
 - Smaller patients require less sweep, consider adding CO₂
 - First gas should be slightly acidotic 7.25-7.35, PaO₂ 80-100mm/Hg
- Appropriate flows goals
 - 100-150 ml/kg/min
 - MAP: appropriate for weight and age
 - Cardiac index: 2.0-2.5L/min/m²
 - SvO₂: >60%
 - Decreasing serum lactate level

Post-Cannulation Stabilization

- Ensure adequate ECMO flow: usually ~ 100-150 mL/kg/min
 - More flow may be needed in patients with aortopulmonary runoff (i.e., through a systemic to pulmonary shunt)
- Adjust flow to maintain adequate tissue perfusion (see below and routine management)
- Evaluate tissue perfusion and assess for end-organ dysfunction
 - Vital signs and hemodynamics
 - Laboratory studies
 - Lactate, arterial and venous blood gases, SvO₂
 - Liver function tests
 - Renal function tests
 - Hematologic and anticoagulation studies
 - Obtain CXR or ultrasound to evaluate cannula position
 - Assess limb perfusion with peripheral cannulation and consider distal perfusion cannula if indicated
- Monitor for and treat bleeding
 - Correct abnormal hematological values
 - Assess for cardiac tamponade in postoperative patients
- Evaluate for LA hypertension and need for LV decompression in patients with LV failure
 - CXR and echocardiogram
 - If LA hypertension is present, consider
 - Inotropes and afterload reduction to augment ventricular ejection
 - LA vent or creation of atrial septal defect via transcatheter approach

Routine Management

- Cardiovascular
 - Monitor vital signs and tissue perfusion
 - Maintain adequate mean arterial blood pressure (MAP)
 - ~35-45 mmHg for neonates depending on size
 - ~ > 60-70 mmHg in pediatric/adult patients depending on age and size
 - Maintain adequate preload and have volume readily available
 - Minimize SVR which may inhibit ECMO flow and tissue perfusion
 - Avoid excessive hypothermia
 - Provide adequate analgesia and sedation
 - Consider use of afterload reducing agents such as milrinone and nitroprusside
- Respiratory
 - Mechanical ventilatory support is typically minimized
 - In VA ECMO ventilator settings should be adjusted and individualized per patient to maintain lung volume, minimize alveolar collapse,

- provide adequate pulmonary venous saturation and coronary oxygenation
 - Avoid increased PVR and consider
 - Pulmonary vasodilator therapy (i.e., iNO)
 - Minimizing pain and agitation
 - Routine ETT suctioning as needed, attempt to minimize trauma to carina
- Neurologic
 - Avoid hyperthermia
 - Assess level of consciousness, pupil response and signs of seizure activity
 - Consider EEG for suspicion of seizures, particularly if muscle relaxed
 - Routine head ultrasound in infants to assess for intraventricular hemorrhage
- Analgesia and sedation
 - Analgesia and sedation as appropriate to minimize oxygen consumption
 - Avoid routine pharmacological paralysis if possible to assess neurological status and promote spontaneous respirations
 - Consider daily drug holiday to assess neurological status if on muscle relaxants
 - Routine assessment of pain and sedation with developmentally appropriate tools
- Infection
 - Usual clinical indicators of sepsis may be unreliable while on ECMO
 - CBC
 - CRP
 - Consider antibiotic prophylaxis
- Fluid and nutrition
 - Manage fluid overload and promote patient diuresis
 - Routine diuretics as appropriate
 - Ultrafiltration and hemodialysis if necessary
 - Routine evaluation of electrolytes and renal function tests
 - Consider parenteral nutrition, initiate enteral feeds when possible
 - GI prophylaxis
- Integument and positioning
 - Change position every two hours as appropriate, caution needed to avoid cannula kinking or dislodgement
 - Routine assessment for pressure ulcers
 - Maintain body alignment to avoid peripheral neuropathy
 - Routine assessment of distal perfusion with femoral cannulation
 - Physical therapy as indicated
- ECMO complications (vary by age, ECMO indication and duration of support)
 - Hematologic
 - Bleeding at cannulation sites, surgical sites, or visceral
 - Clotting events: patient or circuit

- Neurological injury
 - Stroke
 - Intracranial hemorrhage
 - Seizures
- Infection
- Mechanical failure
- Renal dysfunction

Anticoagulation for ECMO

General Principles

- Effective anticoagulation is essential for successful management of ECMO
- Bleeding and thrombosis (circuit or patient) are common events during ECMO
- Age related differences in the coagulation pathway are important considerations in pediatric ECMO

Unfractionated Heparin (UFH)

- The most commonly used anticoagulant for ECMO
- Mechanism of action
 - Interaction of a unique polysaccharide sequence present in 1/3 of heparin molecules with antithrombin III (AT3)
 - Heparin-AT complex then inactivates thrombin, Factors IIa, Xa, IXa, XIa and XIIa
- Pharmacokinetics
 - Varies by patient weight
 - $\frac{1}{2}$ life is approximately 1–2 hours
 - Heparin is cleared largely by renal mechanisms
 - Clearance is faster in neonates than in older children
 - Effect and duration of effect increase with larger doses
- Heparin dosing and titration for ECMO
 - Bolus dose of 50–100 units/kg (max 5000units) at ECMO cannulation
 - Maintenance infusion is titrated based on anticoagulation testing

Common Anticoagulation Lab Tests for ECMO

- Tests used to assess anticoagulation effect of heparin vary widely
- Frequency of anticoagulation testing should be based on type of test used and should be determined by institutional protocol (see Table 2)
- Reduced AT 3 levels can affect the therapeutic action of UFH.
- Monitoring and if necessary repletion should be considered
 - Normal AT3 levels vary by age, but are generally in the 80–120%.
 - Neonates tend to have lower AT levels
 - Replacing AT3
 - FFP: 10–20ml/kg Q8–12 hour until desired AT3 level
 - Human AT (Thrombate III): bolus dosing
 - $\frac{[(\text{desired level\%} - \text{measured AT3 level\%})]}{1.4} \times \text{weight (kg)}$
 - Repeat doses to achieve desired levels
 - Recombinant AT (Atryn): load and maintenance
 - Loading: $\frac{[(100\% - \text{AT 3 level\%})]}{1.3} \times \text{weight (kg)}$
 - Maintenance dose/hour: $\frac{(100\% - \text{AT 3 level\%})}{5.4} \times \text{weight (kg)}$
 - Titrate infusion to maintain desired range

Test	ACT	aPTT	Anti-Xa	TEG
Type	Whole blood clotting	Intrinsic Pathway	Heparin Activity	Whole blood clotting/fibrinolysis/platelet function
Sample Type	POCT	Lab test	Lab test	POCT/lab test
Normal Values	80-160 sec	30-40sec	—	—
ECMO Values	180-240 sec	1.5x normal	0.35-0.71IU/mL	No documented goal values for ECMO but widely used to trend and assess the patient's ability to bleed and clot

Table 2. Common Anticoagulation Assays Used in Managing Patients on ECMO

Other Agents for Anticoagulation in ECMO

- Heparin alternatives may be considered for patients with Heparin induced thrombocytopenia (HIT)
- Direct thrombin inhibitors including Argatroban and Bivalirudin have been successfully used in ECMO patients with HIT
 - Argatroban
 - IV bolus: 100 µg/kg (range: 75–200) over 3-5 min
 - IV maintenance: 2 µg/kg/min (range: 0.1 – 12)
 - Monitor ACT (target 180 – 220 seconds) or aPTT 1.5 – 2.5 X normal
 - Impaired clearance in patients with hepatic dysfunction
 - Bivalirudin
 - IV initial dose: 0.15–0.2 mg/kg/hour with additional IV bolus doses 0.1–0.5 mg/kg can be given to achieve goal aPTT
 - Monitor aPTT with goal of 1.5–2.5 X baseline
 - Impaired clearance in renal dysfunction

Managing Bleeding on ECMO

- Bleeding is common especially in postoperative patients and may require reduction or temporary discontinuation of heparin infusion
- Blood products should be administered to correct underlying coagulopathy
- Antifibrinolytic agents and Factor VIIa may be useful to manage surgical bleeding
 - Epsilon aminocaproic acid (Amicar)
 - Bolus: 100mg/kg (maximum 5000 mg) IV
 - Infusion: 30mg/kg/hour (maximum 1250mg/hour)
 - Discontinue after 72 hours or when bleeding has stopped
 - Adjust dose in patients with renal dysfunction
 - Tranexemic Acid
 - Bolus: 100 mg/kg IV

- Infusion at 10mg/kg/hour for 1–2 hours
- Adjust dose in renal dysfunction
- Factor VIIa (Novoseven): 90 µg/kg IV
 - May be repeated every 2 hours until hemostasis achieved
 - There is a high risk of circuit failure, thus administration is reserved for refractory bleeding

Strategies to Prevent Thrombosis

- Optimize anticoagulation
- Monitor ECMO circuit for early signs of thrombosis and increase anticoagulation to prevent propagation of clot
- Consider increasing anticoagulation during low flow states (i.e., weaning)
- A new ECMO circuit should be readily available to manage acute clot induced circuit failure for patients who require discontinuation of heparin or use of pro-coagulant agents

Readiness for ECMO Weaning

Cardiac Assessment

- Prior to weaning, it is imperative to consider the etiology that led to ECMO support and whether further investigation and potential interventions, including cardiac catheterization or reoperation, are needed
- Consider echocardiogram to assess ventricular function, residual lesions, impact of medications and interventions
- In postoperative patients, further evaluation with cardiac catheterization should be considered to rule out residual lesions if unable to wean from ECMO in 48-72 hours
- Optimize hemodynamic function with vasoactive support
 - Establish adequate heart rate and AV synchrony: if not in sinus rhythm, consider pacing
 - Optimize ventricular loading conditions and consider inotropic support

Pulmonary Assessment

- Evaluate pulmonary compliance
- Use PEEP to maintain adequate end expiratory lung volume
- Assess oxygenation (i.e., A-a gradient, oxygenation index)
- Assess ventilatory capacity by weaning ventilator support
- Drain pleural effusions if necessary

Weaning ECMO Flow

- Monitor central venous pressure, intracardiac pressures, systemic blood pressures, pulse pressure, and systemic perfusion as flow is weaned
- Monitoring trends in NIRS (cerebral and renal) is often helpful
- Venous oximetry and serum lactate are useful to evaluate tissue perfusion
- Perform an echocardiogram when the flow has been weaned to low flow (20-30mL/kg/min) or if there is any deterioration in hemodynamics during weaning
- If patient is stable on low flow ECMO, use bridge between the arterial and venous cannula and allow the patient to be isolated from the ECMO circuit (i.e., clamped)
- If the patient is stable, as determined by continuous monitoring of hemodynamic and laboratory parameters, consider decannulation in 1-2 hours after isolating the ECMO flow

Failure to Wean from ECMO

- Review the patient's cardiac and respiratory function to determine if the patient's status could be further optimized
- If patient fails several weaning attempts despite cardiopulmonary optimization and exclusion of residual defects, then consider whether the patient is a candidate for heart transplant and if so, consider conversion to a ventricular assist device

Extracorporeal Cardiopulmonary Resuscitation (ECPR)

Introduction

- ECMO is a potentially lifesaving intervention for patients who are unresponsive to traditional cardiopulmonary resuscitation (CPR) efforts
- Cardiac arrest is defined as the need for chest compressions or direct, open chest cardiac massage
- American Heart Association (AHA) guidelines for CPR recommend consideration of ECMO to aid cardiopulmonary resuscitation in patients who have a reversible event and good quality CPR
- Established protocols and personnel essential to achieve good outcomes
- Contraindications to ECMO should apply to ECPR patients (see contraindications section)

Initiation and Management

- Rapid initiation of ECPR may be indicated for unsuccessful CPR
- Requires coordination of operative team, ICU team and ECMO team, to successfully cannulate during a cardiac arrest
- Veno-arterial (VA) ECMO is used in all cases of ECPR
- Vascular access: thoracic or peripheral VA ECMO cannulation may be used at the discretion of the surgical and medical teams
- Management
 - CNS protection during and after CPR is critical, hyperthermia should be avoided
 - Once cannulation is achieved ECMO management should continue as for all other ECMO cases (see patient management section)
- Favorable outcomes are reported in many single center studies
 - These range from 30-75% survival to hospital discharge
 - The Extracorporeal Life Support Organization (ELSO) registry reports the use of ECPR in children is increasing and indicates a survival of 39%

ECMO in Infants with Shunt-Dependent Circulation

- Indications
 - Preoperative support is rarely required
 - Inability to separate from CPB
 - Acute hypoxemia (includes shunt thrombosis)
 - Progressive low cardiac output syndrome (LCOS) in the immediate postoperative period
 - ECMO to support cardiopulmonary resuscitation (ECPR)
- Special considerations
 - There may be significant run-off through the systemic to pulmonary artery shunt
 - High ECMO flow (as high as 150-200 mL/kg/min) may be required to maintain adequate systemic blood flow and pressure.
 - Mechanical ventilatory support and sweep gas should be adjusted to maintain adequate gas exchange
 - Supplemental CO₂ may be administered to increase PVR and prevent respiratory alkalosis
 - Occasionally, surgical clips may need to be placed on the systemic to pulmonary artery shunt to reduce the effective diameter of the shunt if systemic oxygen delivery is compromised. However, complete occlusion of the shunt is generally not recommended as doing so has been associated with pulmonary infarction
 - The patient's own lungs can occasionally be used for ventilation and oxygenation, the membrane can be isolated and the circuit used as a VAD (no membrane oxygenator ECMO)
 - An attempt to isolate the membrane may be made within 24 hours of ECMO support
 - Causes for inability to isolate the membrane include an occluded shunt, arterial cannula position in close proximity to the shunt or severe lung disease
 - The membrane cannot be isolated when the source of pulmonary blood flow is a Sano shunt

ECMO after Superior Cavopulmonary Connection (SCPC) and Fontan

- Pulmonary blood flow is passive with both the SCPC and Fontan circulation and this poses unique challenges with initiation and maintenance of ECMO support
- Indications
 - Failure to separate from CPB
 - ECPR, though outcomes are poor
 - Failing Fontan before takedown to SCPC, Fontan revision or as a bridge to transplantation or VAD placement.
 - Progressive LCOS
 - Respiratory failure

- Special considerations
 - Prior knowledge and documentation of venous anatomy (example bilateral SVC or right or left sided SVC) and patency of vessels (both veins and arteries) is important
 - Early and adequate decompression of the SVC is important to maintain adequate cerebral perfusion particularly with the SCPC
 - Multiple venous cannulae may be necessary to achieve adequate drainage of the Fontan circulation (i.e., combination of internal jugular and femoral vein or chest and femoral vein)
 - Venous cannula may need to be upsized particularly in older children and adults with Fontan circulation to achieve adequate systemic venous drainage
 - Investigate early for reversible causes especially with acute Fontan failure in the immediate postoperative period
 - Patients with cavopulmonary connections are at risk for poor neurological and overall outcomes following ECPR for several reasons, including
 - Maintaining adequate end organ perfusion during the resuscitation and cannulation are difficult in the setting of high cerebral venous pressure
 - Central venous anatomy may be abnormal
 - Significant hypoxemia may be present prior to cannulation

Section 2

Ventricular Assist Devices (VAD)

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Introduction

- VAD support may be used as bridge to recovery or transplant
- The Berlin Heart EXCOR was approved for use in children by the FDA in 2011
- Children present unique challenges due to variation in size, anatomy and physiology

Indications

- Acute decompensated congestive heart failure refractory to conventional medical therapy; in some instances, ECMO may provide easier and more rapid deployment
 - Acute myocarditis
 - Post cardiectomy ventricular failure
 - Intractable arrhythmias
- Chronic end-stage heart failure refractory to medical therapy
 - Cardiomyopathy
 - Ventricular dysfunction following surgery for congenital and acquired heart defects

Relative Contraindications

- Ongoing sepsis
- Significant pulmonary disease (consider ECMO)
- Uncontrolled coagulopathy
- Severe neurologic injury
- Irreversible renal or liver failure
- Significant aortic regurgitation can interfere with forward flow, but can be overcome by oversewing the aortic valve
- Prosthetic mitral valve may become frozen shut and is often removed at the time of LVAD placement

Types of Devices

- The two broad categories are pulsatile or non-pulsatile (axial or centrifugal)
- Pulsatile devices (Thoratec PVAD, Berlin Heart EXCOR and Syncardia TAH-t) are paracorporeal devices with a fixed stroke volume
 - Berlin Heart EXCOR is the most commonly used pediatric device
 - In pump diastole, blood is drawn into the device by negative pressure, during systole blood is ejected from the device
 - All patients have a palpable peripheral pulse

- Non-pulsatile devices (Heartmate II and Heartware) are intracorporeal and provide continuous forward flow
 - Non-pulsatile devices have largely replaced pulsatile devices in adults
 - These devices provide continuous flow using centrifugal or impellar-based mechanics
 - Peripheral pulses are not palpable in these patients

Short-term vs. Long-term devices

- Short-term devices: ECMO, Tandem Heart, Impella
- Long-term devices: Berlin Heart EXCOR, Thoratec PVAD, Syncardia TAH-t, Heartmate II, and Heartware

Management Principles after VAD Placement

- Output from the VAD is dependent on preload and afterload
- It is important to appreciate that RV output is preload to the LVAD
- Unloading of the LV by the LVAD has generally made the need for BiVAD uncommon
 - Leftward displacement of the ventricular septum into the LV may compromise LV filling
 - RV may not tolerate the increase in systemic venous return
 - RV afterload may remain elevated due to long standing pulmonary venous hypertension
- Assess for RV failure by monitoring CVP, echocardiogram and cardiac output
- Consider administering iNO or other pulmonary vasodilators in the immediate postoperative period and consider inotropy to improve RV function
- Causes of inadequate filling of LVAD include
 - Low intravascular volume
 - Tamponade
 - RV dysfunction (decrease forward flow and ventricular interdependence)
 - Pulmonary hypertension
 - Arrhythmias
 - Device malfunction
- Neurologic assessment as indicated
- Monitor and manage surgical bleeding, anticoagulation practice varies depending on device
- Wound and device exit site care and management per device protocol

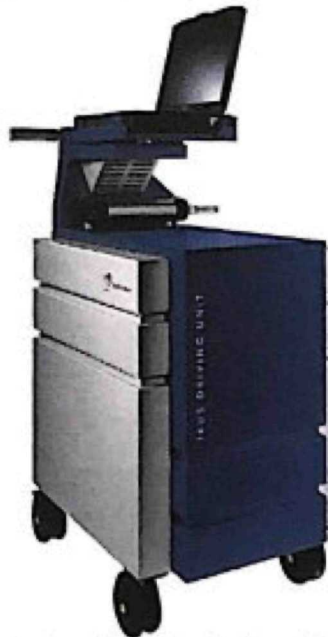
Complications

- Common complications include bleeding, thromboembolic events, neurologic injury, sepsis and multiorgan failure
- Other common adverse events include pancreatitis and arrhythmias
- Persistent arrhythmias can affect RV performance and warrant antiarrhythmic therapy
- Device malfunction

Berlin Heart EXCOR

Overview of Device

- Berlin EXCOR is an extracorporeal, pulsatile, pneumatically driven VAD
- Only FDA approved device for infants and children
- Can be used for biventricular support (BiVAD) or LV support only (LVAD)
- Components include inflow and outflow cannulae, pump, driver (IKUS) and connectors/tubes
- The IKUS controls pump rate, systolic and diastolic pump pressures and % (time in) systole



Berlin Heart permission obtained from Berlin Heart, The Woodlands, Texas

- Pump sizes available: 10 mL, 15 mL, 25 mL, 30 mL, 50 mL, 60 mL



Patient Selection

- Indications and contraindications (see above)
- Appropriate device size based on patient weight
- FDA approved for patients 3-60 kg and 0-18 years old

Device Specific Postoperative Management

- See introduction for details regarding management in the postoperative period
- Management
 - Routine postoperative care (see Management Principles After VAD placement above)
 - Pump checks to assess filling, ejection, and fibrin deposits/clots
 - Wean cardiorespiratory support as tolerated
 - Treat arrhythmias
 - Follow wound care guidelines
 - Perioperative antibiotic coverage per institutional practice
 - For isolated LVAD, assess RV function and output and consider RV afterload reducing agents and inotropy
- Pump troubleshooting
 - Poor filling
 - RV failure
 - Pulmonary hypertension
 - Tamponade
 - Low preload
 - Excessive bleeding
 - Mechanical obstruction
 - Oversized pump
 - Poor ejection
 - High afterload
 - Outflow cannula obstruction
 - Inadequate time in systole
 - Persistent low cardiac output in spite of optimal medical management
 - Consider increasing the pump rate provided good filling/ejection
 - Consider upsizing the pump if the ventricle is not adequately decompressed at full pump capacity (good filling/ejection, low cardiac output)
- Anticoagulation/anti-platelet therapy
 - May follow the Edmonton Anticoagulation and Platelet Inhibition Protocol for Berlin Heart
 - Start anticoagulation with UFH at 24 hours postoperative if
 - Platelets > 20,000/ μ l
 - No active surgical bleeding (no more than 2ml/kg/day)
 - Normal coagulation profile and platelet function
 - Unfractionated heparin (UFH)
 - Starting doses: 15 units/kg/hour (for <12 months of age) or 10 units/kg/hour (for >12 months)

- Titrate to anti-Xa assay for a goal of 0.35 to 0.7 IU/ml as per the Edmonton Anticoagulation and Antiplatelet Protocol for Berlin Heart
- Monitor PT, PTT, anti-Xa and AT3 levels
- May transition to low molecular weight heparin or warfarin when stable therapeutic levels achieved on heparin and patient is stable
- Low molecular weight heparin (LMWH)
 - Transition to LMWH if
 - 2-4 days postoperative
 - Normal creatinine
 - No acute bleeding
 - Starting doses: (turn heparin off and start LMWH simultaneously)
 - Patients $\leq$ 3 months: 1.8 mg/kg Q12 subQ
 - Patients >3-12 months: 1.4mg/kg Q12 subQ
 - Patients 1-5 years: 1.2mg/kg Q12 subQ
 - Patients 6-18 years: 1.1mg/kg Q12 subQ
 - Goal LMWH level/anti-Xa assay: 0.6-1 IU/mL, obtain 4 hours after the second dose
- Warfarin
 - Transition to warfarin if
 - Patient is greater than 12 months of age
 - No acute bleeding
 - Hemodynamically stable
 - Stable enteral feeding regimen
 - Pre-INR obtained (INR can be elevated due to recent cardiopulmonary bypass)
 - Starting dose: 0.2mg/kg, do not exceed max loading dose of 5mg/day (consider 0.1mg/kg for Fontan patients or patients with poor liver function)
 - Bridge to warfarin with heparin until goal INR is achieved, simultaneous administration of warfarin and heparin is indicated (approximately 4 days)
 - Once goal INR is achieved (2.7-3.5), heparin/LMWH can be discontinued
 - If INR falls to 2-2.7: start LMWH dose 0.5 mg/kg (target LMWH level of 0.35-0.5 IU/mL)
 - If INR falls INR <2: start LMWH dose 1 mg/kg (target LMWH level of 0.5-1 IU/mL)
 - If unable to achieve a stable INR with warfarin consider using LMWH
- Anti-platelet agents (aspirin +/- dipyridamole)
 - Aspirin 1 mg/kg divided every 12 hours (or 0.5mg/kg Q12hours)
 - Goal arachadonic acid (AA) inhibition:70-95% via TEG with platelet mapping (see below)
 - Start 4-7 days post implantation and once all drainage tubes are removed
 - Dipyridamole 4 mg/kg divided every 6 hours

- Goal ADP Net G: 4-8 via TEG and platelet mapping
- Start 48 hours after surgery with no acute bleeding
- Clots or fibrin strands
 - Monitor and document any fibrin deposits and clots
 - Treatment: optimize anticoagulation/anti-platelet therapy
 - Ensure complete pump ejection
 - Operate pump at highest tolerated rate that permits optimal ejection and filling
 - Monitor pump head more frequently in the setting of fever, bleeding, or inadequate ejection
 - Indications for pump change: clots >5mm, multi-color, or mobile clots
 - Early neuroimaging if any focal neurologic signs are present (device is not MRI safe)

Overview of Device

- Continuous flow, centrifugal non-pulsatile device
- Magnetically levitated and hydrodynamically-suspended wide blade impeller acts as the only moving part to generate flow
- Thin layer of blood between impeller and pump housing, no points of mechanical contact
- Implanted into the ventricle and housed within the pericardial space
- LVAD configuration with inflow implanted into LV apex and outflow graft anastomosed end-to-side to ascending aorta
- RVAD and BiVAD support possible with device inflow cannula positioned in patient's right ventricle or right atrium and outflow graft connected to patient's pulmonary artery
- Driveline externalized percutaneously and connects the HVAD pump to controller and power source



- Controller connected to two power sources at all times (two lithium ion batteries or AC/DC power adaptor) and programmed by monitor
- Monitor displays
 - Alarms
 - Calculated flow (liters per minute)
 - Measured power consumption (Watts)
 - Programmed operated speed (revolutions per minute)
- Flows reported by controller are calculated, taking into account the power consumption, operating speed and inputted hematocrit. If hematocrit is incorrect, may get falsely low or high calculated flows
- Pump speed (revolutions per minute, RPM)
 - Capable of 1800-4000 (optimal 2400-3200)
- Parameters monitored
 - Flow (cardiac output in L/min)
 - Calculated not measured (based on speed, hematocrit and power)
 - Flow rate up to 10 liters per minute

- Flow is preload-driven and afterload-sensitive
- Power (Watts)
 - Measured number (2.5-16)

Patient Selection

- Patient size selection dependent on
 - Size of ventricle (device inflow 25 mm, therefore requires ~ 30 mm clearance within ventricular cavity)
 - Size of patient thoracic cage
 - Recommend CT chest 3-dimensional imaging to assess for device fit in small patients (<17 kg)
 - May need to alter implantation strategy (diaphragmatic surface) for smaller patients

Device Specific Postoperative Management

- Close attention to preload and afterload
 - Potential causes of decreased preload (low device flows)
 - Right heart failure
 - Tamponade
 - Hypovolemia
 - Changes in intrathoracic pressure
 - Increased PVR
 - Potential causes of increased afterload (low device flows)
 - Pulmonary or systemic hypertension
 - Need to update hematocrit in monitor to ensure accurate reported flows
- For LVAD patients assess RV function and output
- Pump troubleshooting
 - Low flow
 - If waveform amplitude small (<2 L/min), then consider
 - Hypovolemia: consider giving volume, holding diuretics, or decreasing the RPM to allow more filling of the LV
 - Hypotension: check MAP using doppler
 - Consider tamponade if respiratory variation in waveform (pulsus paradoxus)
 - If waveform amplitude large >4 L/min, then consider
 - Fluid overload or inadequate unloading of the ventricle: check fluid balance, check daily weight, consider diuresis
 - Hypertension: VAD is afterload sensitive and will not be able to provide forward flow against higher resistance, therefore more blood will be left in the ventricle and waveform will be wider (recommend MAP <75mmHg)
 - Normal flow but high amplitude waveform
 - Can have very high waveform amplitude with no change in average flow

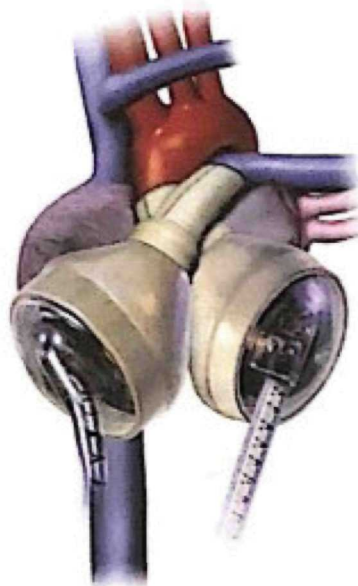
- Need to update hematocrit in monitor to ensure accurate reported flows
- If waveform amplitude >4 L/min, then consider retrograde flow from the ascending aorta through outflow graft into pump and out the LV apex inflow cannula. To treat increase RPM in increments of 100 until trough of waveform is above 2 L/min and waveform amplitude between 2-4 L/min.
- High flow
 - Concern for pump thrombosis or inflow/outflow occlusion in the setting of high power
 - Need to update hematocrit in monitor to ensure accurate reported flows
- Waveform variability
 - Determine if the peak of the waveform varies with each beat or every few beats
 - Consider ectopic beats or arrhythmia (evaluate telemetry)
- Suction events: grouped waveform variability
 - Suction occurs when the cannula is too closely opposed to septum, mitral valve tissue, or LV free wall
 - Can occur in the setting
 - Hypovolemia
 - RV failure
 - Bradycardia (RV is dependent on HR)
 - Poor cannula positioning (with cannula directed to septum, LV free wall or too close to LV tissue)
- Anticoagulation/anti-platelet therapy and management
 - Initiation: 24 hours post-implantation if
 - Platelets $> 20,000/\mu\text{l}$
 - No active surgical bleeding (no more than 2mL/kg/day)
 - Normal coagulation profile and platelet function
 - Unfractionated heparin (UFH)
 - Starting dose: 10 units/kg/hour
 - Titrate to anti-Xa assay for a goal of 0.35 to 0.7 IU/mL based on institutional algorithm
 - Monitor PT, PTT, anti-Xa, and AT3 levels
 - Warfarin: consider transitioning to warfarin if
 - No acute bleeding
 - Hemodynamically stable
 - Stable or progressing enteral feeding regimen
 - Pre-INR obtained (note: INR can be elevated due to recent cardiopulmonary bypass)
 - Starting dose: 0.2 mg/kg, do not exceed max loading dose of 5mg/day
 - Consider 0.1 mg/kg for Fontan patients or patients with poor liver function

- Goal INR is 2-3 or 2.5-3.5 if patient is smaller and set on lower flows
- Dose adjust per patient's clinical picture and institutional algorithm
- Low molecular weight heparin (LMWH)
 - Typically used for a short-term bridge if patient's INR drops below 1.6 while on the HeartWare VAD or is exceptionally difficult to manage on UFH
 - Starting doses: 1 mg/kg BID; if adult size, could consider 30 mg BID prophylactic dosing
 - If checking levels, goal should be 0.5-1 IU/mL (standard risk of bleeding) or 0.4-0.6 IU/mL (high risk of bleeding)
 - Dose adjustments based on institutional algorithms
- Aspirin
 - 4-7 days post implantation (recommend to start post extubation and once patient is mobilizing as high risk of bleeding in the immediate postoperative period)
 - Ideally receiving some enteral nutrition
 - Starting dose: 1mg/kg divided every 12 hours
 - Goal AA inhibition is 70-95% via TEG with platelet mapping

Syncardia (Total Artificial Heart)

Overview of Device

- Syncardia is a pneumatically driven pulsatile system
- The 70 ml device has been FDA approved in adults since 2004 and the 50 ml device was recently approved
- Use in children is limited to case reports
- The native ventricles are replaced by two semi-rigid polyurethane chambers of 70 ml volume
- The blood is separated from the pneumatic chamber by a flexible, multi-layered, polyurethane diaphragm
- Quick-connect inflow sewing cuffs attach the artificial ventricles to the remaining atria
- 27 mm mechanical valves are at the atria-ventricular junction
- Quick-connect outflow sewing grafts attach the artificial ventricles to the pulmonary artery (PA) and aorta
- 25 mm mechanical valves are at the ventricle to aorta and PA
- Percutaneous conduits from the pneumatic chamber attach with the external driver and console. The conduits exit the skin at the upper quadrant of the abdomen
- Pressurized air is shuttered in and out of the pneumatic chambers in synchronization through the attached percutaneous drivelines
- When the sub-diaphragm ventricular pressures exceed the pulmonary and aortic pressures, the 25 mm outlet valves open. The diaphragms will displace upward until they reach maximum distension and all ventricular blood is ejected
- The accumulated pressurized air inside the ventricles is then released back along the drivelines to the console for the beginning of the diastolic filling cycle. Then the 27 mm inlet valves open to allow blood to displace the diaphragms downward
- Systole is indicated by the right sided waveforms on the device monitor. The eject flag on the right sided tracing indicates adequate control of afterload and fill volumes
- Diastole is shown in left sided waveforms on the monitor. The left sided tracing indicates volume status
- Syncardia has the shortest blood flow path for any assist device, with decreased risk of thromboembolism
- RV function is inconsequential
- Arrhythmias are inconsequential



Syncardia permission obtained from SynCardia Systems, Inc. Tucson, AZ

Patient Selection

- Pediatric application of this device is limited by a minimum patient body surface area (BSA) of 1.7 m^2 and antero-posterior chest diameter of 10 cm as assessed by CT scan of the chest

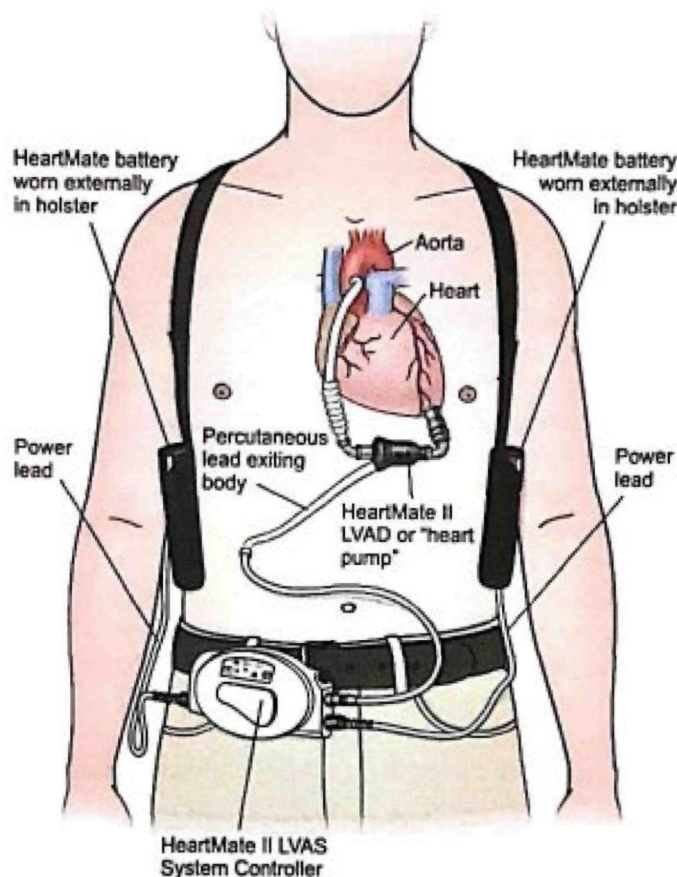
Device Specific Postoperative Management

- Pulsatile perfusion
- TAH patients will not have a heart rhythm
- Typical settings
 - Percentage systole: 50%
 - Heart rate: 120 to 130 beat per minute
 - Left Drive pressure: 180-210
 - Right Drive pressure: 60-110
 - Filling volume: 50-60 mL
 - Typical CO: 7 L/minute
- An echocardiogram may be useful to demonstrate unobstructed systemic and pulmonary venous return
- With increases in intravascular volume, the fill volumes increase which will lead to a decrease in ejection from the device
- Increased SVR may decrease ejection (indicated by the loss of the eject flag) and reduce output. May require afterload reduction
- Attention is given to early detection of tamponade (from IVC and pulmonary vein compression) High CVP with low fill volume indicates tamponade
- Placement of a new central line should only be done if the position of the tip of the catheter can be confirmed to be in the SVC or IVC, and not within the device

- Manage driveline exit sites per institution protocols
- Transition to Freedom Driver when ready
- Anticoagulation management
 - Baseline PT, PTT, platelet count, platelet aggregation studies, TEG and fibrinogen
 - Initiate anticoagulation with heparin after ensuring hemostasis usually over the first 24 hours. Target anti-Xa level of 0.3-0.7 IU/mL
 - Transition to warfarin once there is stable anticoagulation with heparin
 - INR goal 2.5 to 3
 - Once stable, follow INR levels once a week until discharge
 - TEG with platelet mapping to guide anti-platelet therapy
 - Dipyridamole is started first, usually after the first 48 hours when platelet counts are above 40,000 and TEG MA > 56. Target parameters are ADP NET G = 4-8
 - Aspirin is initiated on day 4-5 when TEG platelet mapping AA_{inhibition} < 70%. Target parameters: TEG platelet mapping AA_{inhibition} >70%.

Overview of Device

- Most widely used implantable axial flow device worldwide
- Currently approved as bridge to transplant and destination therapy in adults
- Implanted beneath the diaphragm in either the pre-peritoneal or intra-abdominal location
- Pump
 - Titanium impeller accelerated by magnetic field from the motor producing rotary motion and torque directing blood flow along the axial plane
 - Impeller spins on bearings located at either end of the pump
 - Pump is connected to inlet cannula that inserts into the LV apex and outflow graft that inserts into the ascending aorta
- System controller
 - Main pump connects via driveline to external interface, which controls monitors and provides power to the device
 - Contains backup battery with ~15 minutes of support during power loss
 - May be connected to System Monitor to give detailed display of speed, power, flow, and pulsatility index; and allows for easy access to make adjustments in speed
 - Single power line is small and flexible
 - The portable driver is small and lightweight and thus easily worn on a belt
- Power system
 - Power Module contains backup battery and connects to an AC electrical outlet
 - In portable mode the device is powered by two 14 volt lithium-ion batteries
 - Two power cables connect the batteries or power module to the controller
 - Fully charged batteries power the system for 10-12 hours based on settings



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- Device physiology
 - Operates in parallel with the LV, such that either can supply blood to the aorta
 - Capable of generating blood flow from 3-10 L/minute
 - Power, flow, and pulsatility depend of patient's physiological status.
 - Performance is sensitive to changes in afterload and preload
- Pump speed
 - Only programmed variable
 - Optimal speed is set during "ramped speed study" using echocardiogram in the OR to assess position of the septum, LV size, and aortic valve opening
- Pump flow
 - Calculated based on speed and power
 - Flow across the device is dependent on speed and pressure differential
 - Calculated flows can be inaccurate at speeds <8,000 rpm or <3 L/min and do not account for native LV ejection across the aortic valve

- Pump power
 - Direct measure of current and voltage applied to the motor
 - Influenced by viscosity as well as any degree of inlet or outlet obstruction.
 - Flow obstruction without contact with the rotor will reduce power
 - Thrombus in contact with the rotor will increase power
- Pulsatility
 - Continuous flow device producing non pulsatile flow
 - Pulsatility index is calculated as maximal and minimal flow difference divided by the average flow per cardiac cycle in a 15-second interval
 - Pulsatility index is greatly influenced by LV contractility, intravascular volume, preload, afterload, and by the pump speed

Patient Selection

- Suggested BSA >1.5 m²
- Patients with incompetent aortic valve are poor candidates

Device Specific Postoperative Management

- Hemodynamics
 - Assess RV function and support RV as needed
 - A normal mean arterial blood pressure is targeted as increases in SVR and blood pressure decrease output
 - Pump speed adjustments are based on septal position, LV cavity size, and aortic valve flow with goal ejection every 2nd or 3rd beat
 - Pump speed should not be adjusted to achieve a desired arterial blood pressure
 - Non-invasive BP is most accurately assessed using BP cuff and Doppler
- Hypovolemia
 - Low pulsatility index and suction events in presence of low CVP
 - Confirm with bedside echo evaluating LV dynamics and treat with volume as needed
- Flow obstruction and tamponade
 - Low output with suction events are also suggestive of tamponade physiology in early postoperative phase and should be treated promptly
 - Device flow obstruction leads to elevated LA/LV pressures
 - Physical device obstruction should be addressed surgically
- Anticoagulation
 - Adequate hemostasis should be achieved before anticoagulation is started
 - PT/PTT, INR, and TEG should be assessed daily
 - LDH and plasma free hemoglobin monitored for evidence of hemolysis
 - Aspirin dosing can be based on platelet function tests and clinical condition
 - Use of dipyridamole is dependent on individual center

- Anticoagulation recommendations
 - Initiate UFH 12-24 hour postoperatively based on presence of postoperative bleeding
 - PTT goal 1.2-1.4 times control for 24 hours
 - PTT goal 1.4-1.7 times control after 24 hours
 - Initiate aspirin at 81-325mg daily and dipyridamole 75mg TID on postoperative day 2-3
 - Initiate warfarin (overlapping with heparin) on postoperative day 3-5 and after drains are removed with goal INR 2.0-3.0
 - Maintain on aspirin and warfarin for duration of support
- Anticoagulation regimen should be adjusted if the LVAD flows are low (<3 L/min) or in patients with previous history of device thrombosis

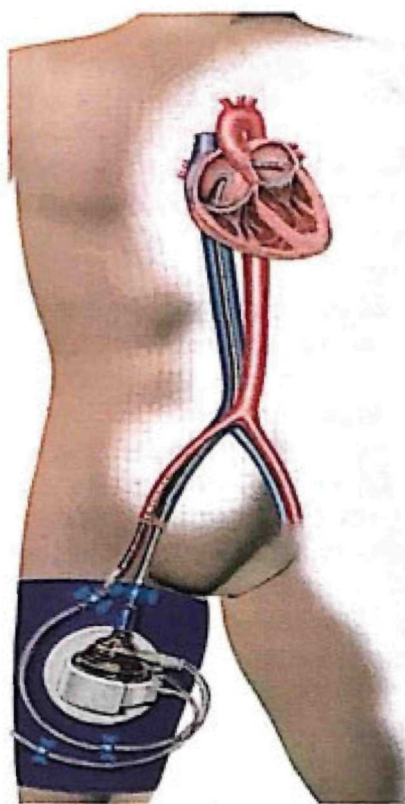
TandemHeart VAD

Overview of the Device

- Short-term or temporary extracorporeal percutaneous LVAD
- Cannulas are inserted percutaneously into the femoral vessels and are attached to the centrifugal pump
- Pump and cannulas
 - A 21Fr. trans-septal cannula, inserted percutaneously through the femoral vein is guided into the left atrium under fluoroscopy
 - The trans-septal cannula (inflow) drains oxygenated blood from the LA to the centrifugal pump
 - Blood is pumped through the arterial cannula/cannulae (outflow) inserted in one or both femoral arteries (15 Fr. or 17 Fr.)
 - An infusion solution (normal saline or heparinized saline at 10 mL/hour) provides a hydrodynamic bearing to the pump
 - In addition, the infusion solution provides anticoagulation to the blood that enters the pump and cools the motor
- **Escort Controller/Console**
 - Provides the following information
 - Cardiac output flow can deliver up to 5 L/min
 - Speed of the centrifugal pump (RPM), maximum speed 7500 RPM
 - Pressure of the infusion solution in pressure transducer (200 – 500 mmHg)
 - Pressure of infusion solution provides power to the pump
 - Contains self-diagnostic abilities and detailed alarms for continuous user observation
 - Built in batteries that provide up to 60 minutes of power.
 - Back-up controller- dual redundant motor control units provide safety

Patient Selection

- Body weight > 30 kg
- The TandemHeart System may be used for temporary extracorporeal circulatory support in older children or adolescents
- Examples include fulminant myocarditis, post-cardiac surgery heart failure or post-transplant rejection
- Contraindications
 - Patients who require biventricular or long-term support
 - Left atrial thrombosis



Tandem Heart permission obtained from CardiacAssist, Inc

Device Specific Postoperative Management

- Pump filling
 - As indicated by the pump flow (L/min)
 - Infusion solution pressure can increase or decrease depending on pump flow/filling
 - Unusual chattering or chugging of the cannulae or pump will indicate inadequate pump filling
- Cannula dislodgement
 - Trans-septal venous cannula position: if dislodgement is suspected, assess for the following
 - Change in color of blood in pump from bright red to dark red due to decreased blood oxygen saturation
 - Dislocation detected by echocardiogram or chest x-ray
 - Arterial cannula position: if dislodgement is suspected, assess for the following
 - Excessive bleeding from insertion site
 - Formation of a hematoma at the insertion site (may be from a tear or dissection)
 - Low flow alarm (measured flow less than 1 L/min and associated hemodynamic instability)
 - Displacement from original skin marking

- Assess for decreased limb perfusion in extremity with arterial cannula.
 - If perfusion is compromised a distal arterial sheath can be placed to improve blood flow and perfusion
- The infusion assembly tubing on the pump should not be routinely changed due to risk of introducing air into the system
- Dextrose will damage the pump and should not be used for the infusion solution
- Do not use acetone in the room of a patient with a Tandem Heart as it will degrade the cannulas
- During travel or AC power failure, built in batteries provide the pump with up to 1 hour of uninterrupted operation. To ensure the batteries are fully charged, the controller must always be plugged in
- Verify the pump is secure in the leg holster and away from patient's skin to prevent burning. If the patient is too small to attach the pump to, position the pump in the bed away from the patient's skin
- Do not cover the pump with bed linens
- Emergency care
 - Defibrillation can be utilized if indicated without risk of damaging the cannulas or pump
 - If the decision to stop the pump is made, the outflow tubing to the patient should be clamped
 - The pump should not be stopped for more than 5 minutes without the consideration of additional heparin
 - Performing CPR can potentially result in damage to the intra-cardiac structures and/or cannula dislodgement. The pump may remain running during the CPR process
- Anticoagulation
 - ACT or PTT used for monitoring anticoagulation.
 - ACT should be 400 seconds during insertion.
 - ACT should be maintained >200 seconds during support period.
 - PTT should be maintained between 2.5 to 3 times normal range during support period

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